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NEOPLASMS OF VASCULAR ORIGIN IN THE MEDIASTINUM

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PRIMARY neoplasms of vascular origin in the mediastinum are rare. This article reviews the cases so far reported in the literature, and reports 3 new cases which present some interesting features.

INCIDENCE

Seybold, McDonald, Clagett, and Harrington (1949) made the first collective review of these tumours, in which they mainly discussed their pathology. They could find only 14 cases, to which they added 3 more cases from the Mayo Clinic.

The real incidence of these neoplasms in relation to other mediastinal masses is very difficult to estimate. However, the widening scope of thoracic surgery, together with the increasing use of mass radiological surveys, has led to the discovery and treatment of many such cases. Altogether, 66 cases have been mentioned in the literature. Of these, tumours originating in the pericardium (9 cases) and the cases belonging to the hæmangiopericytoma group (3 cases) are discussed separately for convenience. Of the remaining 54 cases, only 34 are included for review in *Table I*, together with the 3 cases here described, these being the cases which have been reported in sufficient detail. Twenty-one other cases, which were briefly described or referred to by different writers while reviewing their series of mediastinal tumours, are enumerated in *Table II*. The cases described by Watson and Diamond (1947) and Schorr, Braun, and Isaac (1954) were excluded from *Table I* because of doubt about their final diagnoses in the authors' reports.

Age Incidence.—No age seems to be exempt; the youngest patient was a premature infant (Doderlein, 1938) and the oldest was a 61-year-old patient (Hosoi and Stewart, 1931). Only 6 cases were over the age of 40 years, but it is of significance to note that 6 cases were below one year of age.

Sex Incidence.—Both sexes seem to be equally affected, 20 cases being in males and 17 in females.

Incidence of Malignancy.—Ten cases were found to be malignant and 25 were benign. In 2 cases the tumour was reported as arousing suspicions of malignancy. The highest incidence of malignancy was in the third and fourth decades where 7 out of 14 cases were malignant.

Location in the Mediastinum.—The group of vascular tumours is stated to occur in all parts of the mediastinum with equal frequency (Seybold and others, 1949). However, in the cases reviewed in the present study, it was found that the anterior mediastinum was much more frequently affected (26 cases), more noticeably so in benign cases. The tumour not uncommonly reaches such considerable dimensions as to make it difficult to describe it as located in one part or the other. Most masses bulge radiologically to one side of the chest, though occasionally into both pleural cavities. Only in 7 cases was the tumour mainly posteriorly located. Two of the reported tumours originated in the diaphragm muscle.

DIAGNOSIS

It is difficult to establish the correct diagnosis of hæmangioma before thoracotomy is performed. There was only one case in this series (Heuer, 1924) in which the correct pre-operative diagnosis was reached, after histological examination of a needle-aspiration biopsy of a malignant mediastinal tumour that had eroded the ribs posteriorly. In the case reported by Adams and Bloch (1944), hæmangioma was considered the most likely diagnosis when a biopsy of a related mass in the neck was examined. This points to the importance of searching for a cutaneous or subcutaneous swelling, particularly in the cervical region in a patient with a mediastinal shadow of a doubtful nature. Actually, 7 of the reviewed cases had a benign hæmangioma protruding from within the chest. In Stewart's case (1888), a malignant tumour eroded through the sternum producing a pulsating tumour mimicking an aneurysm. In Brindley's case (1949), there were multiple glomus tumours in the right groin and leg, which were of the same nature as the mediastinal tumour. Carlson and Adams (1952) reported a huge posterior chest wall hæmangioma which extended into the posterior mediastinum as well as into the medullary canal producing hemiplegia. Inasmuch as the mediastinal mass in this case was evidently an extension from the chest wall tumour, it was not included in the present series. Owing to the predominantly anterior position in the

Table I.—Review of Cases of Blood-vessel Tumours in the Mediastinum

Author and Year	Sex and Age	Clinical Picture	Radiography	Treatment	Result	Pathology
Stewart, 1888	M. 32	Cough, palpitation 1 yr. Very painful pulsating swelling over sternum 2 mth. Œdema of face and right arm		Rest and iodides for ? aortic aneurysm though negative history of syphilis	Died after developing dysphagia and ankle œdema	Angiosarcoma in anterior mediastinum infiltrating heart, S.V.C., aorta. Metastases in lung
Shennan, 1914	F. 23	Hæmoptysis 5 yr. Multiple hæmorrhages. Enlarged spleen		Deep X-ray treatment over spleen	Died, terminal mesenteric thrombosis	Hæmangioma, benign, in anterior superior mediastinum. Marked proliferative activity. Angioma in skin, lung, spleen, ovarian cysts
Kott, 1922	F. 36	Pain in chest. Dyspnœa, progressing rapidly, 5 mth.	Shadow filling left chest, more intense in 3 mth., displacing heart to right		Died of exhaustion	Angiosarcoma with invasion of anterior chest and metastases
Heuer, 1924	M. 33	Pain in right chest 5 yr. Cough and dyspnœa 1 yr. Loud systolic murmur 'extra-cardiac' over right chest posteriorly	Large mass in right upper chest mainly anterior. Not pulsating	Radium treatment, no response. Thoracotomy, inoperable. Decompression at 6th, 7th, 8th ribs posterior. Tumour pulsating	Died 4 mth. later	Malignant pleural endothelioma
Heuer, 1924	F. 17	Swelling below left scapula, 8 mth., became larger and painful. Pulsating and systolic bruit over it. Aspirate biopsy showed endothelioma or sarcoma	Mass left lower posterior mediastinum. Destruction of 9th and 10th ribs	Excision of mass and pleura and ribs over it	Well after 2 yr.	Endothelioma (pleural). No metastases malignant but pleura over tumour showed plaques and masses of tumour
Lilienthal, 1927	F. 4	Lump medial to right scapula when 8 mth. old. Recent paresis of both legs progressing to flaccid paralysis	Mass in right lower chest extending to left above the heart. Partial erosion of adjacent ribs and vertebræ. Mass present when 8 mth. old	Evisceration of tumour without opening the pleura. Treated with Cooley's fluid	Well 10 yr. after. X-ray chest: Normal paraplegia persisted	Angiosarcoma, highly malignant invasive properties
Brustalon and Parere, 1927	F. 51	Cough, pain in chest, increasing. Firm pulsating mass in right supraclavicular region. Hæmoptysis. Hoarseness of voice	Mass in right superior mediastinum anterior, pushing trachea to the left		Died before treatment	Endothelioma, malignant. Metastases on diaphragm markedly proliferative
Winkelbauer, 1929	M. 29	Pain in right chest. Slight dyspnœa and cough	Mass in right chest anteriorly, pushing heart to left	Excision readily accomplished	Died 2 mth. after operation of recurrence and metastases in both lungs	Tumour excised: cavernous hæmangioma, benign. Post-mortem: Malignant hæmangio-endothelioma
Hosoi and Stewart, 1931	F. 61	Cough for 1⅛ yr. following attack of pneumonia, hoarseness of voice, and sense of choking. Retrosternal dullness across upper chest	Mass in right superior mediastinum behind the œsophagus. Not pulsating. X-ray negative 7 mth. before		Died in hospital before treatment	Malignant endothelioma, encapsulated. No metastases
Pettersen and Romanus, 1933	M. 5 mth.	Lump, band-shaped, over left shoulder and left scapula, bluish colour; grew rapidly in 2 mth. Cough and stridor. Terminal dyspnœa and cyanosis	Upper mediastinum as broad as heart shadow. Mass in anterior. No increase in size in 3 mth.		Died after several weeks in hospital	Cavernous hæmangioma at site of thymus, growing ouside chest wall on left and into neck
Doderlein, 1938	M., prem. 7 mth.	Autopsy findings			Died during delivery	Hæmangioma diffusely affecting diaphragm

Table I.—REVIEW OF CASES OF BLOOD-VESSEL TUMOURS IN THE MEDIASTINUM—*continued*

AUTHOR AND YEAR	SEX AND AGE	CLINICAL PICTURE	RADIOGRAPHY	TREATMENT	RESULT	PATHOLOGY
Duvoir, Picot, Pollet, and Gaultier, 1939	F. 16	Asthmatic symptoms. Hæmoptysis once, not repeated. Congenital macrodactyly, multiple lipomata. Patient syphilitic	Shadow in left hilar region 8 yr. before, gradually increasing in size; filled left lower chest posteriorly	Thoracotomy; lung showed angioma of left lower lobe besides mediastinal angioma	Tumour unchanged after 4 yr. Died of progressive asphyxia	Lipo-angioma in mediastinum. Angiomata of left lung, liver, spleen. Associated lipomata in stomach
Adams and Bloch, 1944	M. 34	Cough persisting after a respiratory infection. Irregular movable mass in neck	Mass in left upper chest anterior. Multiple rounded opacities, more dense peripherally	Excision incomplete	Well 4 yr. later	Cavernous hæmangioma with phleboliths and bone-marrow elements
Bergstrom, 1945	F. 26 d.	Laboured breathing and cyanosis, getting worse 2 wk. Recent cough and vomiting	Right diaphragm obscured by dense triangular shadow at cardiophrenic angle. Mass is anterior. No movement of right diaphragm		Died 2 wk. after onset of symptoms	Benign hæmangioma. Capillary and cavernous replacing muscle of diaphragm
Bergstrom, 1945	F. 36 d.	Black and blue areas of skin on hips and back of hands. Regurgitation of bloodstained mucus			Died about 1 wk. after symptoms	Cavernous hæmangioma involving capsule and intralobar connective tissue of thymus and tissues around
Seybold, McDonald, Clagett, and Harrington, 1949	M. 27	Pain in the chest for 3 wk. Loss of weight of 8 mth. duration	Mass anterior to left lung hilum, pushing trachea and heart to right. Not pulsating. Increased in size during last 4 mth.	Exploration revealed mass filling of mediastinum and invading pericardium. Biopsy only	Developed superior vena caval obstruction, ? cerebral metastases. Died after 2 mth.	Hæmangio-endothelioma grade I malignancy with invasive properties, ? metastases
Seybold and others, 1949	M. 18	Mass radiography showed a shadow in right chest. Pain in lumbar region since age of 10, radiating in last 2 yr. into abdomen. ? Spondylolithesis. Lost 10 lb. in weight	Mass in right chest in relation to D.9–10 vertebræ. Erosion of D.10 vertebra. No change in size in 6 mth.	Excision. Tumour found to invade sympathetic ganglion. Post-operative radiography	Well 3 yr. after treatment	Hæmangio-endothelioma with invasive nature
Seybold and others, 1949	M. 59	Mass radiography finding. No symptoms	Mass well circumscribed posterior to hilum of right lung	Excision	Well 8 yr. after operation (Ellis and others, 1955)	Benign hæmangioma
Thomas and Chesser, 1950	F. 18	Mass radiography showed a shadow in the chest. Vague distress in chest for 1 yr.	Mass in region of left hilum anteriorly	Excision		Cavernous hæmangioma with aggregations of lymphoid cells in stroma
Horst and Beatty, 1951	M. 20	Mass radiography pick-up. Dullness over left base	Mass occupying most of left chest anteriorly. Not pulsating, mediastinum central	Excision difficult in region of the pedicle		Hæmangiofibroma
Hirschfield, 1951	F. 3 mth.	Soft swelling in right supraclavicular region. Respiratory distress, more marked on compressing mass	Shadow across apical part of right chest, displacing mediastinum to left continuous with neck mass	Aspiration of neck cysts, later thoracotomy and aspiration of mediastinal cysts. Post-operative irradiation	Slight improvement. Died 2½ mth. after operation	Hæmangioma of neck and anterior mediastinum with spindle cell matrix
Schlumberger, 1951	M. 26	Incidental autopsy finding				Fibrolipohæmangioma in anterior mediastinum cone
Perasalo, 1952	F. 48	Cough and dyspnœa after ? influenza. Dullness over left lower chest	Shadow in left lower and mid-zones, anterior, well defined, ? pulsating. X-ray 2 yr. before negative	Excision of mass with capsule. Adhesions to lung	Well 1 yr. later	Hæmangioma with suggestion of malignancy
Maggi, Barousse, and Cardeza, 1952	F. 24	Dyspnœa and cough, worse in 2 mth. Dysphagia, cyanosis. Telangiectases on face and anterior chest wall	Bilateral shadow across the mediastinum, lobulated, anterior, more bulging on right side	Treated with nitrogen mustard for a possible lymphoma	Died	Hæmangio-endothelioma, capillary and cavernous

Table I.—REVIEW OF CASES OF BLOOD-VESSEL TUMOURS IN THE MEDIASTINUM—*continued*

AUTHOR AND YEAR	SEX AND AGE	CLINICAL PICTURE	RADIOGRAPHY	TREATMENT	RESULT	PATHOLOGY
Grimes, Raphael, and Stephens, 1953	F. 31	Shadow in mediastinum discovered on routine X-ray 4 yr. before; only slight increase in size recently	Mass well-circumscribed in superior mediastinum posteriorly and on the right	Excision of well-encapsulated but adherent tumour		Cavernous hæmangioma
Sebesteny, 1953	F. 47	Pain in left scapular region increasing in last 5 mth. Shortness of breath	Mass in front and below left hilar region	Excision with capsule with no difficulty		Cavernous hæmangioma
Maurer, 1953	F. 38	Shortness of breath. Pain in the chest	Mass above and anterior to left hilum, shown by angiocardiography to be extravascular	Thoracotomy in prone position inadequate. Sternum-splitting thoracotomy: excision	Well 5 wk. later	Endothelioma in region of great vessels. No signs of malignancy
Keegan, 1953	F. 18	Loss of weight and anorexia. Epigastric pain	Mass anterior to left hilar region; smaller mass below it. Angiography showed pressure on superior vena cava	Thoracotomy: biopsy only. Post-operative irradiation done elsewhere	Condition unchanged 7 mth. later	Benign hæmangioma invading heart and possible invasion of a lymph-node
Emery and Doxiadis, 1953	M. 8	Cough, dyspnœa on slight exertion, and loss of appetite for 3 wk. Hard lump in right side of neck. Prominent veins over right chest. Horner's syndrome	Opacity filling almost all left chest. Mediastinum pushed to right. 4 yr. before there was opacity of left upper zone diagnosed as pneumonia	Following bronchoscopy cough and stridor became so severe as to necessitate tracheotomy	Died 2 hr. after tracheotomy	Benign hæmangioma with compression of the big veins of neck and trachea, which were mainly posterior to tumour
Jenny and Ulsperger, 1954	M. 26	General fatigue. Pain in chest for 5 wk.	Homogenous shadow in right hilar region, smaller rounded shadow in left hilar region. Both anteriorly placed	Excision of right lung and right mass. Excision of left mass, sternal split approach	Died 3 mth. after operation	Malignant hæmangio-endothelioma invading right lung and lymph-node metastases
Valle, 1954	M. 23	Mass radiography finding	Mass in lower anterior mediastinum, bulging to the left side	Excision	Well 18 mth. after	Hæmangioma (benign) adherent to diaphragm and other mediastinal tissues
de Maria, 1954	M. 1 yr. 5 mth.	Bluish compressible mass in left supraclavicular region. Sudden increase in size 1 mth. before on violent coughing	Shadow obscuring upper ⅔ of left lung field, displacing mediastinum to right side	Excision without difficulty. Left upper lobe atelectatic		Hæmolymphangioma extending into the neck
de Maria, 1954	M. 42	Asthenia for 2 mth. Acute attack of cough, expectoration, and pain in the chest	Mass in anterior and middle mediastinum bulging on left side between left upper lobe, bronchus and diaphragm	Excised totally. Found adherent to pericardium and great vessels		Hæmangio-endothelioma, benign in nature
Ellis, Kirklin, and Woolner, 1955	M. 9	Shadow in mediastinum found on routine X-ray. Easy fatigability	Mass in upper zone of left lung field anteriorly confluent with mediastinum, possibly pulsating	Total excision, dissected with difficulty	Well 2 yr. after operation	Cavernous hæmangioma, benign in nature
Balbaa and Chesterman, 1957. Present series: *Case* 1	M. 11	Pain in left chest, transient. Hæmoptysis for few days	Shadow in superior mediastinum bulging into both sides. Obstruction of superior vena cava	1952: thoracotomy and biopsy. 1955: excision incomplete	Well 10 mth. later	Benign hæmangioma. Marked infiltrating edges
Case 2	M. 7 wk.	Difficulty in breathing. Mass in right neck and axilla	Shadow in superior mediastinum, pushing trachea to left	Attempted excision after test irradiation 1000 r.	Died during operation	Hæmolymphangioma
Case 3	M. 21	Pain in left chest, 5 mth. Rapid weight-loss of 25 lb.	Mass bulging posterior to left hilum	Incomplete excision after test dose of 400 r.	Died 17 mth. after operation	Angiosarcoma with infiltration and recurrence

mediastinum of this group of tumours, especially the benign ones, the possibility of a tumour being an angioma ought to be considered whenever a patient presents with an anterior mediastinal shadow.

In 7 cases the tumour was merely an incidental radiological finding in essentially asymptomatic persons. This observation confirms the value of mass radiological surveys in the diagnosis of intrathoracic lesions.

The commonest presenting features were pain in the chest, cough, and dyspnœa, which are equally common in all mediastinal tumours and cysts and are non-specific. Almost all asymptomatic patients had a benign tumour. As previously mentioned, a mass in the axilla, root of the neck, or over the thoracic cage was noted in some cases. Among the rarer symptoms were Horner's syndrome due to pressure on the upper dorsal sympathetic nerve (Emery and Doxiadis, 1953), hæmoptysis due to an associated vascular tumour in the lung or arteriovenous fistula (Shennan, 1914; Duvoir, Picot, Pollet, and Gaultier, 1939), or occurring in the absence of any lung lesion, as in *Case* 1. Hæmatemesis due to associated vascular tumours in the intestines was found in Shennan's and Duvoir and others cases.

Table II.—Cases of Mediastinal Tumour not described in Detail

Author	Year	No. of Cases	Diagnosis
Watson and Diamond	1947	1	Angiofibroma; Schwannoma?
Harrington	1949	10	Endothelioma (5 cases) Hæmangio-endothelioma (5 cases)
Brewer and Dolley	1949	1	Endothelioma
Tutien	1949	1	Angiofibroma
Svanberg	1952	1	Hæmangioma
Gross	1953	1	Hæmangioma
Talman	1953	1	Hæmangioma
Peabody, Strugg, and Rives	1954	1	Hæmangio-endothelioma
Schorr, Braun, and Isaac	1954	1	Hæmangioma? Lymphangioma? Vascular teratoma?
Ellis, Kirklin, and Woolner	1955	2	Angiosarcoma (2 cases)

RADIOLOGICAL FINDINGS

Blood vascular tumours usually appear as well-circumscribed shadows even in some malignant cases. Although more commonly located in the anterior mediastinum, they have been reported in almost all other subdivisions of the mediastinum. In one of Bergstrom's 2 cases (1945), the tumour which diffusely infiltrated and replaced the right hemidiaphragm showed as a triangular shadow in the right cardiophrenic angle. Although this appeared to be fixed to both the diaphragm and the right heart border, it did not move on respiration (Van Alstyne, 1945).

Erosion of related ribs was observed in 2 cases (Heuer, 1924; Lilienthal, 1927), and erosion of the vertebral bodies in 2 cases (Lilienthal, 1927; Seybold and others, 1949). These were all malignant cases.

Fluoroscopy of the patient's chest showed transmitted pulsations in only a few cases. In Perasalo's case (1952) these gave the impression of fluid in the mass. Expansile pulsations were suspected in Ellis, Kirklin, and Woolner's case (1955). On the other hand, masses which were found to be actively pulsating during thoracotomy could not be recognized during fluoroscopic examination.

Angiocardiography proved rather disappointing as it failed to reveal the vascular nature of these tumours in those cases in which it was done. Probably this is due to the fact that the circulation of blood may be rather sluggish in the cavernous tissue of the neoplasms, which may also be partially obliterated by thrombosis. It could, however, reveal extravascular pressure on the cardiac border (Dotter and Steinberg, 1949) or the great vessels as in *Case* 1 (*Fig.* 679), in which it showed almost complete occlusion of the superior vena cava. It must be pointed out that the number of cases in which angiocardiography has been used is still too small to permit any conclusions to be reached.

Tomography and kymography, where used, added very little to the information obtained by standard radiographs and fluoroscopy. Phleboliths in a mediastinal shadow were demonstrated only in a doubtful case of hæmangioma. Possibly they occur more often, but require a special fine technique to be visualized. Their presence, however, indicates merely that the tumour is vascular and not necessarily unresectable; therefore the finding of phleboliths should not obviate thoracotomy as suggested by Schorr and others (1954).

PATHOLOGY

Neoplasms of blood-vessel origin have always been a source of controversy in the literature, concerning their origin, classification, and nomenclature. The basic structure of this group of tumours is composed of tubes, lined by endothelia, which are enclosed within sheaths of connective-tissue fibres of varying density. A simple classification, based on embryological and histological differences, such as that adopted by Stout (1945) has much to recommend it.

The majority of the benign members of this group of tumours is now considered developmental in origin. Whether they arise from sequestrated embryonic unipotent angioblastic tissue, as orginally postulated by Ribbert (Bergstrom, 1945) or from excessive development of more or less normal vessels, as suggested by Ewing (1940) is still disputed. They have been referred to by some pathologists as vascular 'hæmatomas', while the remaining tumours in this group are considered to originate as a true neoplastic process affecting the vessel walls and the supporting connective tissue. The differentiation between them is rendered difficult by the possible occurrence in the first type of disorderly richly cellular areas, which may be poorly differentiated, particularly in infants. Besides, the vessels may mingle with the surrounding tissues, giving the deceptive appearance of infiltration (Willis, 1952).

Stout (1945) classifies this 'true neoplastic' group into two main types, each of which can be benign or malignant. First, there are the rare but interesting hæmangiopericytoma tumours which arise from the pericytes of Zimmerman, and these will be discussed separately. Second, the hæmangio-endothelioma tumours in which the endothelial cells

play the main role; they are arranged usually in more than one layer around vascular lumina. Excessive proliferation of atypical endothelial cells, together with very freely anastomosing channels characterize the malignant cases. Probably most of the malignant tumours of blood-vessels could be included in this group. Hæmangiopericytoma and hæmangio-endothelioma tumours are sometimes difficult to differentiate and only with the aid of silver reticulin stains and perhaps tissue cultures has this become possible.

Multiple Angiomatous Tumours.—Seybold and others (1949) mentioned as a criterion for inclusion in their series of primary vascular tumours in the mediastinum the absence of co-existent similar tumours in the lung, to exclude the possibility of the mediastinal tumours being metastatic in lymph-nodes. This, however, did not prevent them from including Shennan's case in their series. Willis (1952) states that " . . . while malignant angioblastic neoplasms certainly do occur and sometimes yield blood-borne metastases, the great majority of cases of multiple angiomatous formation are multifocal lesions, frequently developmental in origin ". This can explain the multiplicity of benign tumours in the mediastinum as well as in other sites, e.g., liver, spleen, lung, etc., as described in Shennan's case. This has long been referred to in the literature as a " benign metastatising " hæmangioma, an entity which is denied by Stout (1945) and Ackerman (1953). The fact that the patient was young and had had evidence of the presence of vascular tumours for several years, besides their presence mainly in reticulo-endothelial territories, argues strongly in favour of a multicentric origin. On this basis, the case described by Duvoir and others (1939) can be included in this group of tumours.

Tumour Characteristics.—At thoracotomy, an angioma commonly presents as a well-defined encapsulated soft neoplasm which has a dark plum colour and may be pulsating. But quite often it forms an extensive mass that almost fills an entire hemithorax, while the edges mingle with adjacent mediastinal tissues, giving the deceptive appearance of an infiltrative growth. In some cases it actually strangles some of the main vessels of the mediastinum, e.g., the aorta (Kott, 1922; Bergstrom, 1945) or the superior vena cava (Seybold and others, 1949; *Case* 1 reported here). True invasion of the lumen of the pulmonary artery was described by Shennan (1914). Transgressing the pleural barrier and infiltration of the lung was not noted except in malignant cases. Similarly, erosion of adjacent bony structures is characteristic of malignant tumours.

In some cases the nature of the tumour may not be disclosed until actual dissection of the mass is started or even until histological examination is carried out.

In the case described by Hirschfield (1951), the thin-walled transparent cysts give the impression of a lymphangioma rather than a hæmangioma, but Ewing describes large cystic dilatations filled with clear fluid as consistent with the diagnosis of hæmangioma as long as the microscopic picture is characteristic. On the other hand, some tumours actually show evidence of a mixture of lymphangiomatous as well as hæmangiomatous tissue, hence the term hæmolymphangioma which readily describes the cases of de Maria (1954) and *Case* 2 here reported.

Malignant angiomatous tumours may well be localized and encapsulated and easily shelled out at operation, although usually they tend to invade adjacent organs, erode related bones, and produce lymphatic and even blood-borne metastases. These mostly start as malignant neoplasms, but the question of malignant transformation of benign angiomata seems to be a matter of controversy. Some writers mention its occurrence as a definite hazard; Perasalo (1952) describes in his case what appears to be a recent malignant change in a tumour in which the infiltrative growth is early. Pack and Miller (1950) in their review on hæmangioma are of the opinion that malignant change is such a rare occurrence that it does not influence their treatment. On the whole, there is paucity of histological evidence to support either view in the literature reviewed.

Hæmangioma of the Pericardium.—This forms a small subgroup, of which only 9 cases were reported. Of these, only the case reported by Bencini (1936) was included in the series studied by Seybold and others (1949). These tumours were discussed in a comprehensive review by Greenberg and Angrist (1948). *Table III* includes those cases reported so far in the literature. Mention is made here only of a few of the features which may puzzle the clinician.

The patient presenting a picture of acute cardiac tamponade without a history of trauma may have an intrapericardial rupture of a pericardial angioma

Table III.—Cases of Hæmangioma of the Pericardium

Author	Year	Pathological Diagnosis
Redtenbacher	1889	Angiosarcoma
Lefas	1898	Cavernous hæmangioma
Timme	1915	Cavernous hæmangioma
Bencini	1936	Angioma
Scheideggar	1937	Malignant hæmangioblastoma
Greenberg and Angrist	1948	Cavernous angioma Malignant angio-endothelioma
Stout	1949	Benign hæmangiopericytoma
De Carlo and Lindquist	1950	Hæmangiosarcoma

(Lefas, 1898). This tendency to spontaneous rupture may be accounted for by the constant traumatization inflicted by the beating heart.

Similarly, a previously healthy individual showing unexplained rapid decompensation and venous congestion with an enlarged cardiac shadow should suggest the remote possibility of a gradually progressive bleeding from a pericardial tumour (Redtenbacher's case, 1889). In malignant cases, the increasing bulk of the tumour itself may insidiously produce a comparable picture. Small benign angiomas, however, may remain silent, and be discovered only at routine autopsy. (Bencini's case, 1936.)

Hæmangiopericytoma and Glomus Tumour.—Of this very interesting group of tumours, only 2 cases have been described in the mediastinum, by Brindley (1949), and Ferguson, Clagett, and McDonald (1954). A third case of hæmangiopericytoma in the pericardium was described by Stout (1949). Pathologically the tumour may be

soft and vascular or hard and sclerotic. It affects any age and may even be found in the newborn. It has no characteristic site in the mediastinum and may attain any size. The single characteristic feature of these tumours is the histological picture which essentially shows profuse proliferation of capillaries and endothelial buds, each of which is surrounded by a connective-tissue sheath and one or more layers of concentrically arranged pericytes of Zimmerman. Non-myelinated nerves and nerve-endings may or may not be present. Whether glomus tumours and hæmangiopericytomas are variants of the same neoplasm or are different entities is still disputed. Ferguson and others, however, conclude that " . . . according to Stout's own terminology glomus tumors might well be haemangiopericytomas with the appearance of glomus-like bodies ".

The clinical picture of Brindley's case is interesting, as the patient suffered from chest pain that had the characteristic paroxysmal nature of peripherally situated glomus tumours. Both cases were excised successfully with disappearance of the patients' symptoms. The literature does not contain any case report of a malignant hæmangiopericytoma of the mediastinum.

TREATMENT

Surgical excision is certainly the treatment of choice in benign cases owing to their possible complications. It must be realized, however, that this may be a tedious procedure because of the infiltrative nature of some of these neoplasms, and substantial blood-loss may necessitate transfusion of several pints of whole blood. The desirability of a generous exposure cannot be over-emphasized; in 2 of the reported cases a second operation was necessary to gain a better access to the tumour. This can be avoided, particularly in cases in which the mass appears to bulge into both thoracic cavities, by utilizing an anterior bilateral approach obtained either by splitting the sternum vertically, or by dividing it transversely in line with a bilateral intercostal incision.

In some cases it may be advantageous to stage the operation to obtain complete excision. In *Case* 1, part of the mass had to be left behind. In a future operation excision of this remaining portion, together with the occluded segment of the superior vena cava, replacing it with a homograft, will probably have to be carried out.

Irradiation therapy is not of much value in benign cases, as the tumour is not radio-sensitive. Besides, the patient being a child in a good percentage of cases, may suffer unnecessary damage inflicted on his growing thoracic cage by radiotherapy.

For the malignant cases, total excision is the only chance for cure, probably more so if combined with radiotherapy. The latter alone may offer the patient with an inoperable growth some palliation, though the results are far from encouraging.

Of the reviewed cases, 20 underwent operative removal of the tumour; 11 of these had complete removal, and 9 had only incomplete excision which was followed in one case by deep X-ray therapy. In 3 cases, only a biopsy was made because the tumour was considered inoperable (Seybold and others' 1949), or because the disease was widespread in the lungs and mediastinum (Duvoir and others, 1939). Heuer (1924) attempted decompression of the chest by excision of the overlying ribs without much success, the patient surviving only four months after operation.

Though malignant tumours usually have a poor prognosis, yet in some cases a combination of excision and radiotherapy (Seybold and others, 1949) or even surgical excision alone (Perasalo, 1952) can lead to two or three years' survival. Radiotherapy was considered rather hazardous to attempt in the latter case owing to the presence of active tuberculosis in the lungs.

It might be of interest to note that the longest survival in the malignant cases was in the case reported by Lilienthal, which was treated by eviscer-ation of the tumour, followed by systemic injection of Cooley's fluid; the patient was reported alive and well with radiological disappearance of the tumour ten years later.

CASE REPORTS

Case 1.—A. L., a male child 11 years old, was admitted to the City General Hospital, Sheffield, on account of a large shadow in the superior mediastinum. He had no complaint on admission, but 10 weeks previously he complained of pain in the chest which was sharp and more noticeable on inspiration. This gradually improved without treatment. Seven weeks later he developed a fever of moderate degree accompanied by mild hæmoptysis which lasted only 5 days. There was history of whooping-cough complicated by pneumonia, scarlet fever, chicken pox, and bronchopneumonia, all before the age of 4.

On Examination.—A pale thin child with slight cyanosis. No lymph-nodes were palpable and there was no clubbing of fingers. The tonsils were enlarged. Pulse and temperature were normal. Chest examination revealed a pigeon-breast deformity and Harrison's sulci. There was fullness over the left infraclavicular fossa with some prominent veins and diminished percussion note over the same area. Few rhonchi were heard over the left base. The heart was normal and the blood-pressure was 110/70. No skin lesions were noted. Physical examination was otherwise normal.

Radiography.—Examination of the chest (*Figs.* 676, 677) showed an opacity confluent with the superior mediastinum and protruding to the right and to the left. It had a well-defined convex margin, and appeared to 'sit' on the cardiac shadow. In the lateral film it appeared to lie in the anterior and middle portions of the superior mediastinum. Fluoroscopy confirmed these findings. The trachea was slightly deviated to the right and slightly narrowed in the coronal plane. There was only transmitted pulsation. The shadow did not move with swallowing and barium swallow showed a normal œsophagus. The mass was assumed to be of thymic origin. Laboratory findings were within normal limits.

At Operation.—(Aug. 7, 1952, Mr. J. T. Chesterman.) Left posterolateral thoracotomy through the bed of the resected fifth rib was made. A large nodular mass of varying consistency occupied the upper mediastinum. Dissection was started by opening the overlying pleura, and freeing of the tumour was attempted but had to be given up after one hour, owing to its adherent and infiltrative nature and its extension into the right chest. No enlarged lymph-glands or metastases were encountered. A biopsy was taken and the chest was closed without drainage. The post-operative course was uneventful, the patient being discharged on Aug. 26, 1952.

HISTOLOGICAL EXAMINATION (*Fig.* 678).—A nodule composed of a network of vascular channels, varying in size from that of a capillary to that of a sinus. Some of these channels were thin-walled, with little more than two endothelial layers separating them. There was some fibrous stroma, intervening in places with occasional stromal hæmorrhages. Scanty foci of lymphocytes and fat cells were present. Diagnosis: Cavernous hæmangioma.

Post-operative radiotherapy of the tumour was suggested but refused by the radiotherapist owing to the insensitive nature of the neoplasm and the danger of the procedure on the bony structure of the thorax.

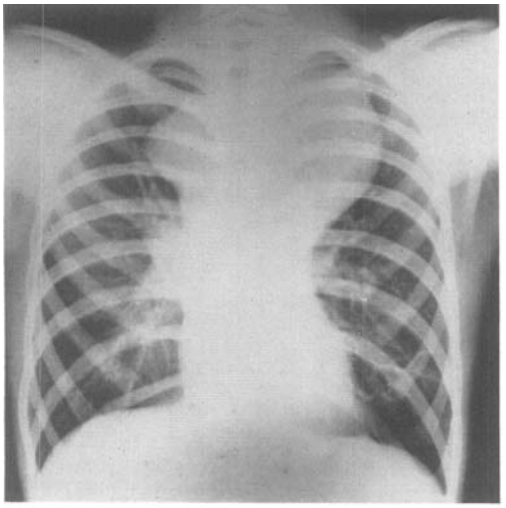

FIG. 676.—*Case* 1. Postero-anterior radiograph.

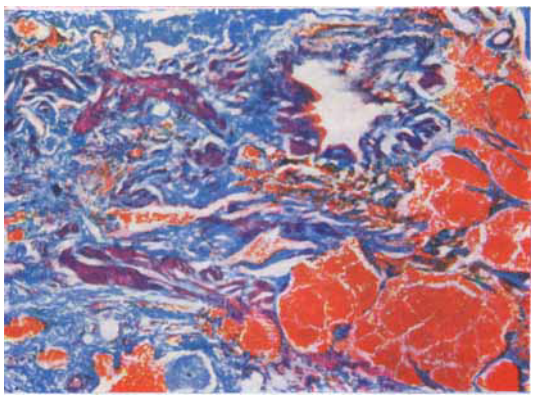

FIG. 678.—*Case* 1. Histological picture. (×50.)

Subsequently, the boy was forced to lead a somewhat restricted life though he could swim and play football a little.

ON RE-ADMISSION.—Owing to some increase in the size of the tumour, local discomfort in the chest, and retarded growth, the boy was re-admitted on March 6, 1955.

ON EXAMINATION.—Congested jugular veins were more noticeable on the right side. Angiocardiography (*Fig.* 679) revealed the presence of almost complete obstruction at the lower end of the superior vena cava; most of the dye escaped through the azygos vein and other collateral channels. The other findings were essentially the same. It was decided to attack the tumour through an adequate exposure.

AT OPERATION.—March 28, 1955. The chest was entered this time through a transverse anterior incision, opening up the right and left second intercostal spaces and dividing the sternum at the same level with a Gigli saw. Excellent exposure was obtained and the tumour was found to extend from beyond the superior vena cava on the right to the region of the descending thoracic aorta on the left. Freeing of the tumour was very tedious, especially the posterior aspect of the central portion, due to the presence of very thin-walled venous channels around the tumour mass. There was a large communicating vessel about 1½ cm. in diameter entering probably into the right atrium, the exact anatomy being very difficult to identify. The greater portion of the tumour and its vascular connexions were excised, but ultimately a portion behind the superior vena cava and the posterior part on the left side had to be left behind when it was realized that the patient's condition would not allow further prolonged dissection. The chest was closed with underwater drainage bilaterally. Seven pints of blood were given, and the operation required about 4½ hours. The patient made a slow but uneventful recovery. Histological examination of the resected specimen showed essentially the same picture as the previous biopsy, with no evidence of malignancy. Ten months later, the patient is developing well and attempting with more success to lead a normal life.

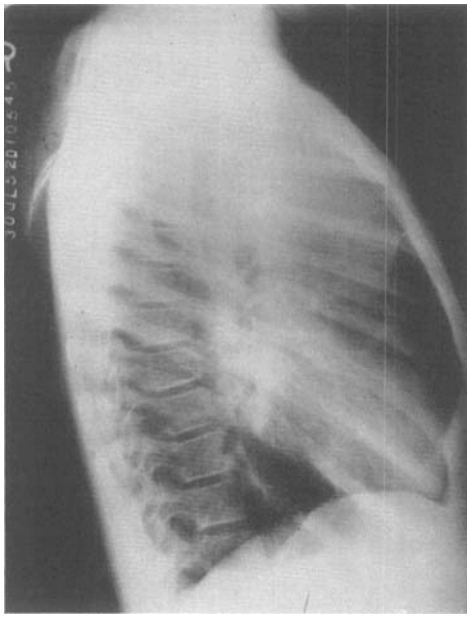

FIG. 677.—*Case* 1. Lateral radiograph.

Case 2.—L. C. B., a male infant 7 weeks old, was admitted to the University of Michigan Hospital in Ann Arbor, Michigan, on June 5, 1940. Difficulty in breathing and choking spells during feeding had occurred during the two weeks prior to admission.

ON EXAMINATION.—A well-developed baby, breathing comfortably when quiet, but showing laboured breathing while crying. The entire right side of the neck was occupied by a large cystic mass which appeared to bulge more on crying, and seemed to extend behind the clavicle into the right axilla. Rhonci were found over both lung fields.

Radiography (*Fig.* 680).—Marked enlargement of the mediastinal shadow above the heart. The trachea was sharply displaced to the left, and in the lateral view it was seen to be narrowed and displaced posteriorly. It was

felt that the mass in the mediastinum was connected to the cervical and axillary masses and it was believed that the lesion was not of thymic origin. Laboratory findings were within normal limits.

Biopsy of the axillary mass revealed a ' lymphangioma cavernosa '. The baby was therefore referred for deep

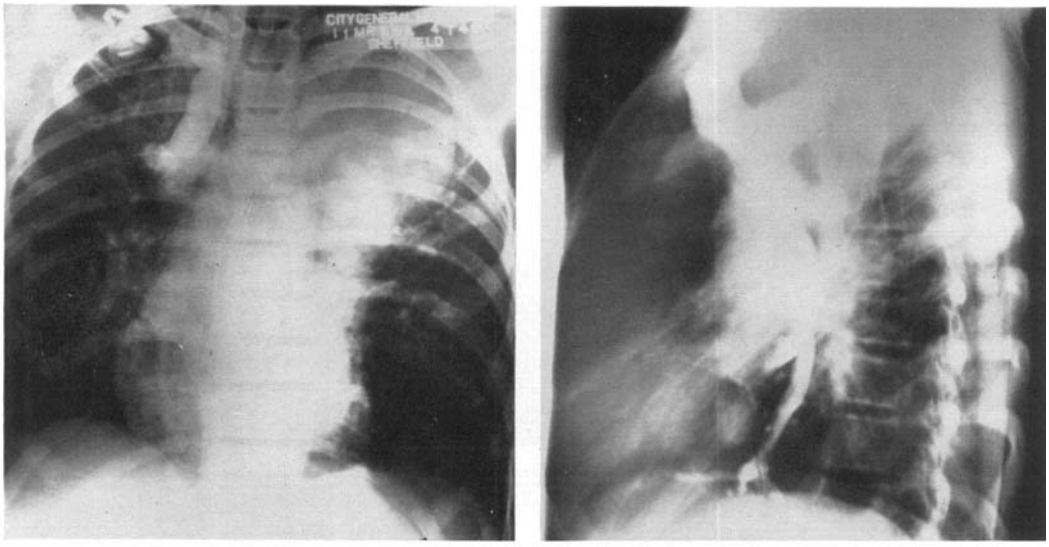

FIG. 679.—*Case* 1. Angiocardiogram showing almost complete occlusion of the superior vena cava, and large accessory vessels connecting the mediastinal veins with the tumour mass.

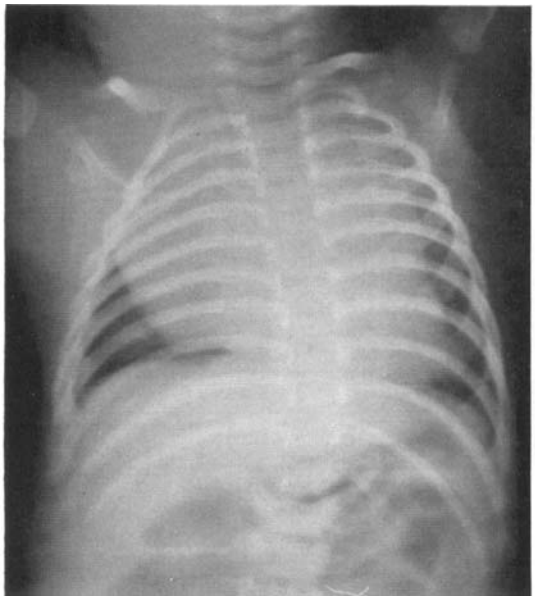

FIG. 680.—*Case* 2. Radiograph showing huge mediastinal tumour. Note the cervical and axillary masses.

FIG. 681.—*Case* 2. Histological picture. (× 100.)

X-ray therapy; 1000 r units at 5 exposures were given to the chest, neck, and axilla. There seemed to be no apparent response to the treatment. Because of rapid deterioration in the baby's condition, an operation was decided upon.

AT OPERATION (June 26, 1940, Dr. John Alexander).—A posterolateral thoracotomy incision was made and the right pleural cavity was entered through the bed of the resected first rib. A large neoplasm was found protruding from the upper mediastinum and displacing the apex of the lung down to the level of the fifth intercostal space posteriorly. It was mostly solid postero-inferiorly and partly cystic anterosuperiorly. Anteriorly it seemed to enclose the superior vena cava and subclavian artery. The tumour was lobulated and adherent to the various mediastinal structures and was infiltrating into the neck and axilla. During attempted excision of the tumour, the infant's condition deteriorated and he died before the operation was completed.

Autopsy revealed considerable narrowing of the trachea, and the cause of death was probably multiple segmental atelectases of both lungs.

HISTOLOGICAL EXAMINATION (*Fig.* 681).—There were areas which consisted of many large endothelial-lined spaces filled with blood-corpuscles. Smooth muscle was present in their walls. Other similar sinusoidal spaces had no blood-cells in them and small collections of lymphocytes were visible between the spaces. The tumour was interpreted as a combination of cavernous hæmangioma and lymphangioma, better named hæmolymphangioma. The tumour in the neck was predominantly a hæmangioma, whereas that in the axilla was mainly a lymphangioma.

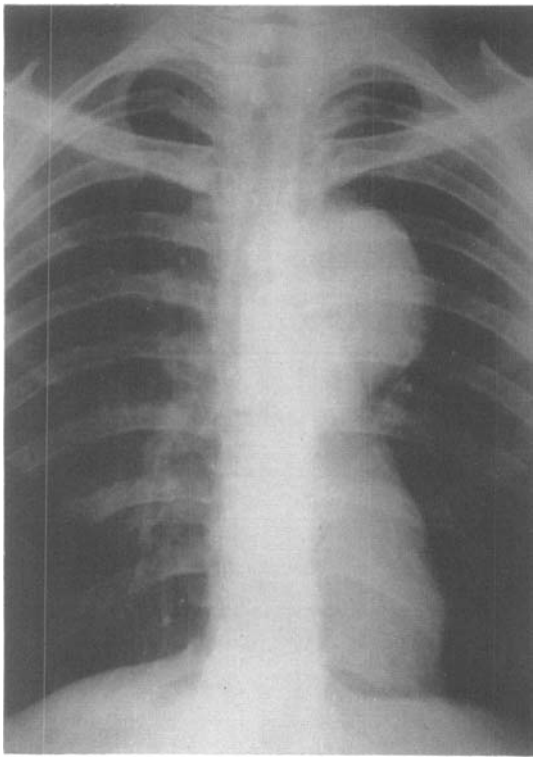

FIG. 682.—*Case* 3. Radiograph of chest.

Case 3.—R. H., a 21-year-old man, was admitted to the University of Michigan Hospital, Ann Arbor, Michigan, on Nov. 3, 1942, with a chief complaint of pain in the posterior portion of his left chest, of 5 months' duration. This started as dull and occasional pain, but became intolerably severe and constant prior to admission. There was a 25-lb. weight loss during that period. A previous history of left pleurisy 2 years previously was obtained. This was associated with hæmoptysis, and this occurred while the patient was doing some work which necessitated exposure to silica.

ON EXAMINATION.—The pulse and temperature were normal. The blood-pressure in the right arm was 105/60 mm. Hg, and 130/90 mm. Hg in the left arm. There was absent tactile vocal fremitus anteriorly and posteriorly over the left upper chest. Breath-sounds had a bronchial quality with scattered râles over this area. There was generalized palpable lymphadenopathy and a large soft gland was palpated high up in the left axilla. Laboratory findings were within normal limits.

Radiography (*Fig.* 682).—A smooth rounded mass bulging into the left thoracic cavity at about the level of the pulmonary hilum, and pushing the trachea slightly over to the right side. In the lateral view, it proved to be posterior to the hilar region, extending toward the vertebræ and ribs, but with no apparent bone erosion. A tentative diagnosis of probable neurogenic tumour or possible lymphoblastoma was made.

A test dose of irradiation of 400 r was delivered in two anterior and two posterior parts, with no significant reduction in the size of the tumour in 6 weeks.

Bronchoscopy on Nov. 6, 1942, showed hyperæmia and œdema of the left main bronchus and compression of the bronchus below the level of the upper lobe orifice.

AT OPERATION.—Thoracotomy was performed by Dr. John Alexander on Nov. 9, 1942, through the bed of the resected fifth rib. A fist-sized tumour was found to bulge into the upper half of the left main fissure and actually invaded the lung tissue. Sharp dissection was required to free it from the lung, vagus nerve, sympathetic trunk, and from the vertebral bodies. Part of the tumour tissue, infiltrating deeply into the anterolateral wall of the thoracic aorta could not be removed, for the aorta was accidentally nicked during the dissection, but was readily sutured. The chest was closed with drainage.

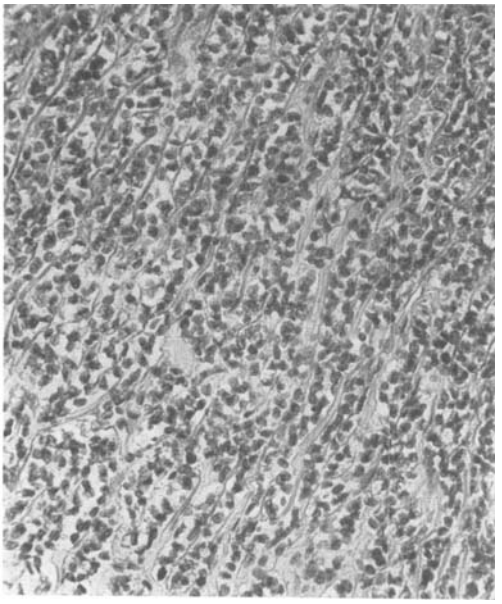

FIG. 683.—*Case* 3. Histological picture. (× 165.)

PATHOLOGICAL EXAMINATION (*Fig.* 683).—A circumscribed and partially encapsulated neoplasm consisting of polyhedral cells arranged in alveoli and columns. In some areas there were obvious vascular spaces lined by neoplastic cells. This was interpreted as an 'angiosarcoma' and the neoplastic cells as of endothelial origin.

The post-operative course was uneventful. Post-operative irradiation of the tumour area with 3000 r was given and the patient was discharged on Dec. 16, 1942. Following discharge, the patient's condition and chest radiographs were satisfactory until November, 1943, when he began to complain of weakness and weight loss and hoarseness of voice. X-rays revealed recurrence of the tumour. His condition gradually deteriorated until he died on March 5, 1944.

SUMMARY

1. The literature concerning the neoplasms of blood vascular origin located in the mediastinum is reviewed. Sixty-six cases were found, but only those reported in sufficient detail are summarized in table form to facilitate future reference.

2. Three previously unreported cases which present some interesting features are reported in detail with photomicrographs.

3. The clinical picture and the value of different diagnostic procedures are discussed in enough detail to help in making a pre-operative diagnosis.

4. Some controversial points in the pathology are discussed.

5. Operative removal is the only effective treatment to avoid the occurrence of possible complications.

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