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CASE REPORT

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Posterior mediastinal hemangioma

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Abstract We report posterior mediastinal hemangiomas in a 4-month-old and a 6-month-old girl. The masses were identified on radiographs of the chest followed by contrast-enhanced CT. Histological evaluation of the surgical specimens established the final diagnosis. Although mediastinal hemangiomas have been described, they remain a rare entity. A diagnosis can be suggested by relatively high attenuating masses on contrast-enhanced CT. Posterior mediastinal hemangiomas sometimes mimic neuroblastomas, which is the most common posterior mediastinal in this age group.

Keywords Posterior mediastinal mass · Hemangioma · CT

Introduction

Hemangiomas are benign, richly vascular tumors that can involute with time. Their occurrence in the mediastinum is rare, with an incidence of less than 0.5% of all mediastinal masses [1]. The posterior mediastinal location, as in our patients, is even rarer. At this site, any mass is considered likely to be a neuroblastoma and has to be distinguished from it. We present a case of a posterior mediastinal hemangioma mimicking neuroblastoma. A second, similar case is presented but with concomitant tumor outside the mediastinum.

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Case reports

Case 1

A 4-month-old full-term girl was diagnosed with tetralogy of Fallot at birth and was scheduled to have it repaired electively. A routine preoperative radiograph of the chest revealed a mass in the right hemithorax (Fig. 1). Contrast-enhanced CT of the chest showed a 2-cm well-defined mass in the posterior right paraspinal region, adjacent to T3 and T4 vertebral bodies (Fig. 2). A portion of this mass extended into the right T3–T4 neural foramen, abutting the dura without causing any significant effacement. The mass was of relatively high attenuation. Given its location and the patient's age, the mass was viewed as a suspected neuroblastoma. There was no adenopathy or any other evidence of metastasis.

The CT also demonstrated a right-sided aortic arch with an aberrant left subclavian artery coursing posterior to the esophagus. A ventricular septal defect was also identified, consistent with the patient's history of tetralogy of Fallot.

A subsequent right thoracotomy revealed a mass in the posterior costovertebral angle extending from the second to the fourth intercostal space and gently compressing the lung forward. The mass was dissected from its extension into the neural foramen and resected in one piece. On gross examination, the mass appeared as a red, soft tissue superficially. Frozen sections suggested hemangioma, which was confirmed by the permanent fixed histological specimens. The patient did well post-operatively. She returned a month later to resume her cardiac care.

Case 2

A 6-month-old girl was transferred from another hospital for management of dilated cardiomyopathy and congestive heart failure. Her work-up included serial

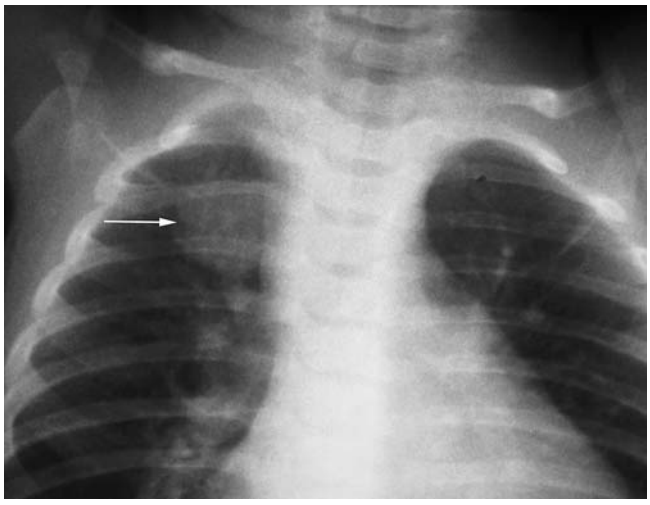

Fig. 1 Plain film of the chest shows a 2-cm, rounded soft-tissue opacity (*arrow*) at the medial right lung apex. There is slight widening of the intercostal space between the right third and fourth posterior ribs, suggesting the location of the mass in the posterior mediastinum

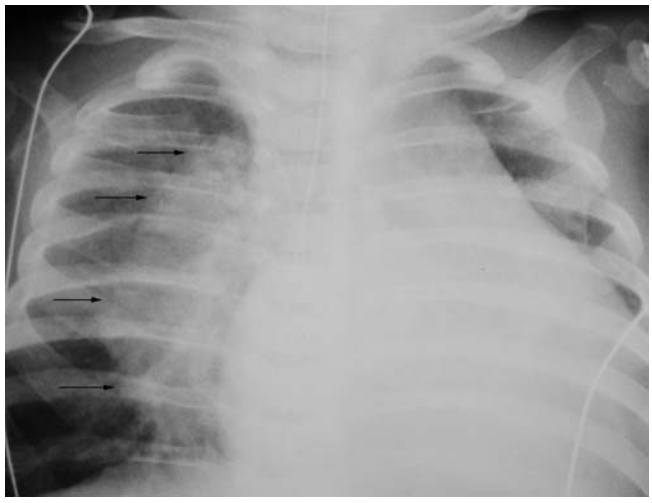

Fig. 3 Radiograph of the chest shows a large mass (*arrows*) on the right. Lack of obscuration of the heart border and slight widening of some posterior intercostal spaces (*asterisks*) suggest posterior location. The heart is moderately enlarged. A nasogastric tube is present

chest radiographs, which demonstrated persistent bilateral airspace opacities related to atelectasis. When the lungs were better aerated, a soft-tissue density was identified in the right posterior paraspinal region (Fig. 3). A contrast-enhanced CT of the chest identified bilateral thoracic paraspinal masses (Fig. 4). These masses appeared to be enhancing. There was extension of the larger mass on the right side, measuring 7.0×2.0×2.5 cm, into the chest wall, beyond the outer rib margin. The left paraspinal mass measured 1.0×1.0×2.0 cm and extended into the neural foramen at the mid-thoracic level. In addition, a large enhancing right suprarenal mass was identified in the upper abdomen (Fig. 4). Because of this finding, additional images were obtained through the abdomen; however, these were delayed relative to the intravenous contrast medium and without benefit of oral contrast medium, yielding a limited evaluation of the abdominal contents. The right suprarenal mass had small central areas of low attenuation and punctate calcifications. There was a compressive effect upon the inferior vena cava. The findings were initially suspected of representing a right adrenal neuroblastoma with metastatic or concomitant disease along the right and left paraspinal regions of the chest and the posterior chest wall. Hemangioma was not strongly considered pre-operatively, so the relationship of the patient's heart failure to the mass was unclear at this time. In addition to the masses, the CT showed cardiomegaly and an incidental finding of a double aortic arch (Fig. 4).

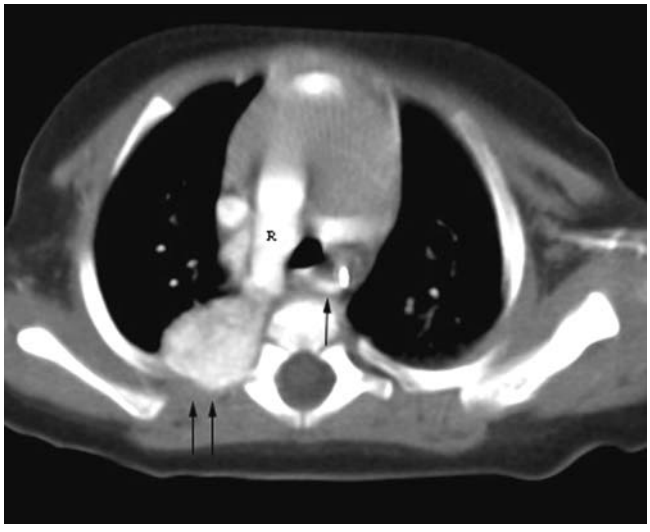

Fig. 2 Contrast-enhanced CT of the chest demonstrates an enhancing mass (*double arrows*) in the posterior right paraspinal region. There is a right aortic arch (*R*) with an aberrant left subclavian artery (*single arrow*)

The patient was taken to surgery, and exploration of the abdomen revealed a multifocal hemangioma. A massive hemangioma of the ileum and cecum was found with two very large feeding arteries and a large draining vein. (This lesion was not readily apparent on the preoperative CT.) This lesion was believed to be hemodynamically significant; therefore, a total of 45 cm of bowel along with the hemangioma was resected. In addition, a large compressible dark red soft-tissue mass was identified in the right upper retroperitoneum. It lay posterior to a normal-appearing adrenal gland and IVC. The appearance was consistent with a hemangioma. It was believed that its anatomic location would make resection in its entirety difficult and would result in a considerable blood loss; hence, it was neither resected nor biopsied. After the surgery, the patient was treated with a dose of 1,000,000 units of alpha interferon. A repeat CT performed prior to discharge showed the right

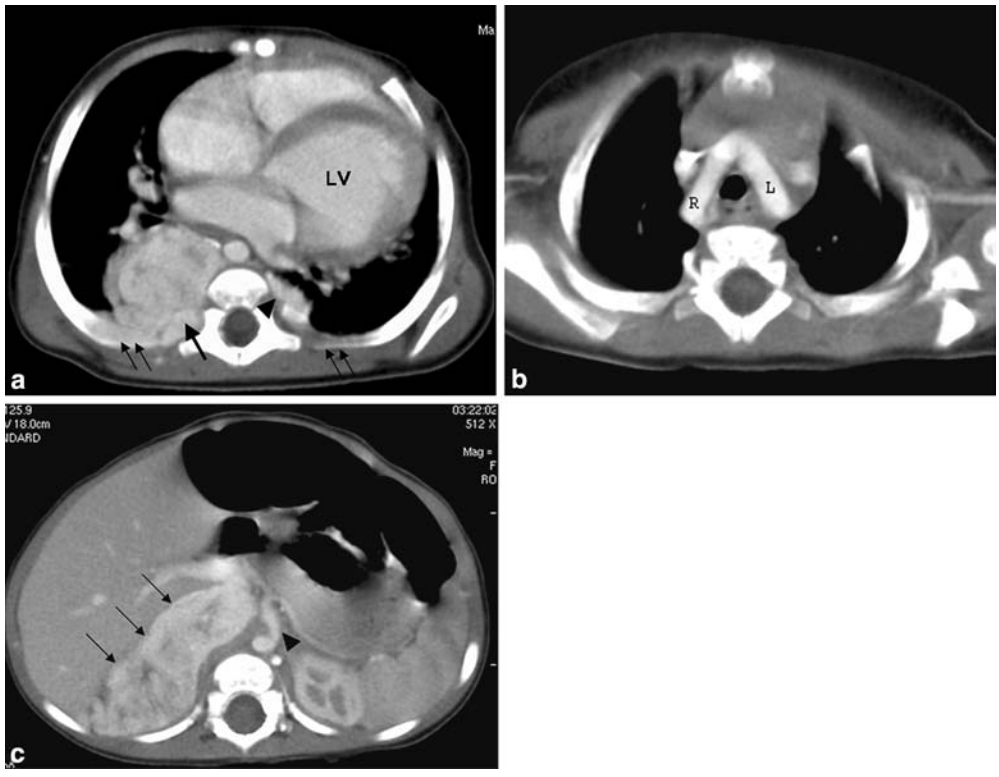

Fig. 4 Contrast-enhanced CT of the chest and abdomen. **a** Bilateral paraspinal masses. The right mass (*arrow*) is larger than the left (*arrowhead*) and extends beyond the outer posterior rib margin. Rib erosions are noted (*small arrows*). The left ventricle (*LV*) is slightly dilated. **b** Double aortic arch, right (*R*) and left (*L*) **c** A large enhancing mass (*arrows*), with central areas of low attenuation, in the right retroperitoneum. Prominence of celiac axis (*arrowhead*) likely reflects high flow with surgically proven mesenteric hemangioma

thoracic and the right suprarenal masses; the left paraspinal mass was no longer seen. Subsequent care was transferred to another institution and further follow-up is not available.

Discussion

A hemangioma is a compact, abnormal collection of blood vessels. It may be found in the skin or in relation to the visceral organs (i.e., lungs, GI tract, eyes, and brain). These benign, vascular neoplasms make up less than 0.5% of all mediastinal masses [1]. According to Taori et al. [2], only 125 well-documented cases of mediastinal hemangioma had been reported in the literature up until 2000. Like other mediastinal masses of childhood, these might manifest by nonspecific symptoms (cough, dyspnea and chest pain) or symptoms related to their anatomic location and extent of invasion. Most often, they remain asymptomatic and are incidentally detected on a routine plain radiograph of the chest.

The vast majority of mediastinal hemangiomas are located in the anterior mediastinum (68%) [2]. The posterior mediastinal location is very rare. Involvement of the middle mediastinum is seen as a continuation of disease from anterior or posterior locations. Most of the posterior mediastinal tumors in children are of neurogenic origin (neurofibroma, neurilemoma, neurenteric cyst, neuroblastoma, ganglioneuroblastoma, pheochromocytoma and neurofibrosarcoma). In these two cases, the location of the hemangiomas simulated tumors of neural crest origin.

On chest radiographs, hemangiomas appear as round or lobulated, well-defined masses. Calcified phleboliths are helpful in suggesting the vascular nature of the mass. However, phleboliths are seen in only 10% of cases [3]. CT is more sensitive in its characterization of the calcifications. The CT appearances of both infantile hemangioendothelioma and adult-type hemangioma in the liver have been thoroughly studied, and imaging protocols have been established [4–6, 7]. Hemangiomas often demonstrate similar properties outside the liver. They appear as homogeneous, low-attenuating masses on a precontrast CT, where phleboliths can be identified, if present. After intravenous iodinated contrast medium administration, there is a characteristic pattern of enhancement that can differentiate these from other mediastinal masses [8]. As in hepatic hemangiomas, delayed images are helpful in displaying slow accumulation of the contrast medium. CT is helpful in evaluating the extent of the tumor and its invasion into adjacent structures.

A posterior mediastinal mass in a young child is most often of neurogenic origin, with neuroblastoma being the diagnosis of import. The avid enhancement of a hemangioma mitigates against thoracic neuroblastoma, which typically enhances much less (Fig. 5) [9]. The smaller lesion in the first patient enhanced homogeneously, whereas the larger lesions in the second patient showed lack of enhancement centrally, likely reflecting delayed central enhancement or partial involution (not pathologically proven). The identification of metastatic disease suggests neuroblastoma; however, metastases are less common with thoracic neuroblastoma, and multi-

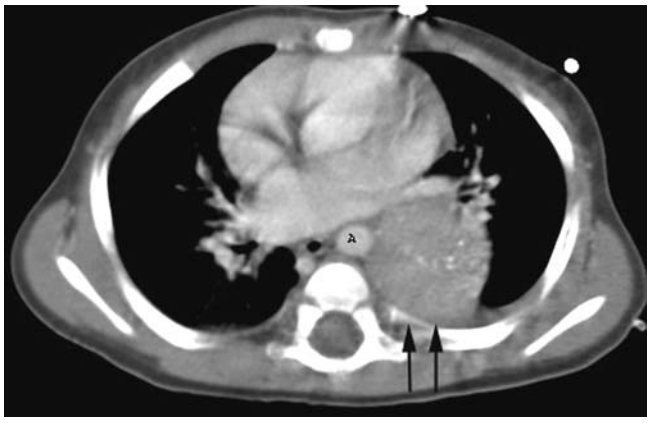

Fig. 5 Contrast-enhanced CT of the chest demonstrates a large mass (*arrows*) in the left posterior mediastinum. This mass has low attenuation and does not appear to be enhancing when compared with the adjacent heart chambers and aorta (*A*). A few punctate calcifications were noted. This was pathologically proven to be a thoracic neuroblastoma

focal hemangiomas can mimic metastatic neuroblastoma (i.e., Case 2). Lack of calcification is not helpful in narrowing the diagnosis, as approximately 90% of these vascular tumors do not exhibit calcification and thoracic neuroblastomas often lack calcification too. MR would probably aid in differentiating a hemangioma from a neurogenic tumor; however, it was not utilized preoperatively in either of our cases. Some neurogenic tumors, namely pheochromocytomas and paragangliomas, might demonstrate MR characteristics similar to those of a hemangioma—hypointense on T1-weighted images and very hyperintense on T2-weighted images [10].

Incidentally, both our patients had accompanying aortic arch anomalies. Of note, the child with tetralogy of Fallot had a right aortic arch with an aberrant left subclavian artery, but not a mirror-image right aortic arch, as is usually seen with tetralogy of Fallot. The association of mediastinal hemangiomas with aortic arch anomalies including coarctation of the aorta and right aortic arch has been reported [11], though the etiology remains unclear. At least two explanations have been proposed for this association: severe isthmal hypoplasia from ischemia results in coarctation caused by increased flow through the hemangioma [12], and low blood output below the coarctation results in low tissue oxygenation and thus enhanced angiogenesis [13]. Neither of these hypotheses seems likely, given the lack of mediastinal hemangiomas in the overwhelming majority of patients with aortic coarctation or aortic arch anomalies.

Mediastinal hemangiomas should be considered an important differential diagnosis of posterior mediastinal masses, even though these are rare. The diagnosis might be suggested by an unusually high attenuation of the mass on contrast-enhanced CT. Suspicion of the diagnosis prior to surgery can greatly reduce the risk of hemorrhage and, in asymptomatic cases, potentially avoid surgical exploration altogether. Further imaging can aid in confirming the diagnosis. Preoperative suspicion of hemangioma, as opposed to neuroblastoma, will likely alter approach and management.

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