RETROPERITONEAL CYSTIC LYMPHANGIOMA

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ABSTRACT — A case of a large retroperitoneal tumor in a previously asymptomatic twenty-two-year-old white female is presented. A review of the literature confirms the rarity of this tumor. Its histologic and embryologic derivation as well as its subtle and bizarre method of presentation are discussed. The cause of such lesions is debatable, but primary cure can be accomplished by meticulous excision of the lesion or marsupialization. This seldom seen neoplasm must enter into the differential diagnosis of all retroperitoneal masses.

A case is reported of a large retroperitoneal cystic lymphangioma. The cause of these neoplasms is speculative, but the most plausible explanation is the possibility that the cysts are due to the continued growth of congenitally misplaced lymphatic tissues. The most frequent symptoms are an abdominal or flank mass. Treatment consists of en bloc excision if possible; however, marsupialization is an alternative. It is a rewarding experience for a surgeon to obtain a surgical cure of this neoplasm, particularly in light of the usual foreboding preoperative diagnosis.

Case Report

A twenty-two-year-old white female was admitted to St. Joseph Mercy Hospital with a two-week history of malaise, lethargy, and daily febrile episodes to 101° F. She also complained of severe and constant pain in the right flank and right lower quadrant. No urinary or gastrointestinal symptoms were noted. Physical examination disclosed an anxious female in moderate distress. Her vital signs were stable, with an oral temperature of 99° F. Abdominal examination revealed a soft, tender, gelatinous mass occupying her right lower quadrant extending across the midline above the umbilicus. Bowel sounds were present but hypoactive, with right flank and costovertebral angle tenderness. Rectal and pelvic examinations were unremarkable.

Laboratory data on admission were as follows: hemoglobin 13 Gm., hematocrit 39, white blood cell count 5,600 per cubic centimeter, with a normal differential. Serum creatinine was 1 mg. and blood urea nitrogen 7 mg. per 100 ml., with normal findings on serum electrolytes and liver function tests. Urinalysis revealed no albumin or sugar, specific gravity was 1.010, pH 5, with 0–1 white blood cells and occasional red blood cells per high-power field with no bacteria. Cytologic examination of the urine on three occasions was negative.

An upper gastrointestinal and small bowel series revealed a large ill-defined radiolucent mass occupying the right flank, displacing the gallbladder, duodenum, and right colon to the left (Fig. 1A). A flattened lower edge to the liver can be seen on the excretory urogram film. Note effacement and extrinsic deformity of right renal pelvocalyceal structures and a gas-filled right colon (Fig. 1B). Abdominal aortogram confirmed an extrinsically deformed nephrogram by the flank mass.[1] Elongated uncoiled right lumbar arteries are shown without evident neovascularization (Fig. 1C). Retroperitoneal liposarcoma was the preoperative diagnosis.

At surgery the right kidney was found to be displaced toward the midline by a large multicystic mass. The mass appeared to be originating from the spine and extending from the inferior

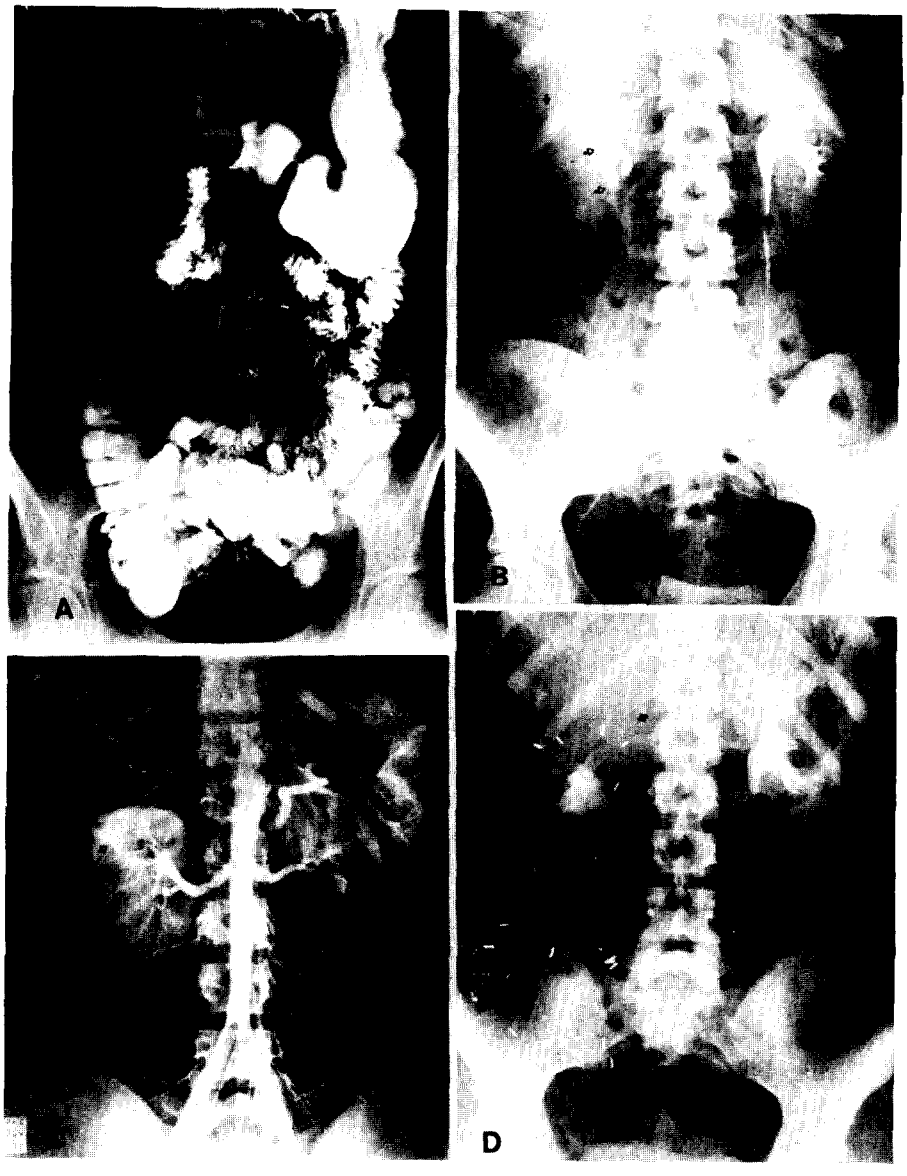

FIGURE 1. *(A) Upper gastrointestinal and small bowel series reveal large radiolucent mass occupying right flank, displacing gallbladder, duodenum, and right colon to left. (B) Excretory urogram noting effacement and extrinsic deformity of right renal pelvocalyceal structures and gas-filled right colon. (C) Elongated uncoiled right lumbar arteries are shown without evident neovascularization on abdominal aortogram. (D) Postoperative urogram illustrating relative normal position of right kidney after excision of neoplasm.*

pole of the kidney to the pelvic brim while incorporating several nerve trunks. Meticulous surgical excision removed the mass in toto while leaving the abdominal and retroperitoneal structures intact. Some minor nerve trunks were sacrificed. The postoperative course was unremarkable with only minimal paresthesia along the distribution of the genitofemoral nerve. Postoperative urogram revealed the relative normal position of the right kidney after excision of the neoplasm (Fig. 1D).

The gross specimen was a 14 by 10 by 4.5 cm., 200-Gm. multicystic mass with cysts measuring to 3 cm. in diameter. The mass was composed of fat and fibrous tissue which was wet and edematous with numerous spaces containing clotted blood and chocolate-colored fluid. Medium-power photomicrographs revealed numerous lymphatic channels lined by endothelium and surrounded by fibromuscular and adipose tissue (Fig. 2). Numerous chronic inflammatory cells infiltrate this stroma.

Comment

The retroperitoneal tissues above and below the kidney may be the seat of a variety of tumors, benign or malignant, cystic or solid. Retroperitoneal lymphangiomas are rare cystic neoplasms of lymphatic origin which resemble cystic hygromas. Saraway in 1893[2] reported the first excision and cure of the neoplasm. Park and Tabah,[3] in their extensive review of 870 primary retroperitoneal tumors, found only 5 to be lymphangiomas. Oviedo, Larson, and Parent[4] have recently found that these cysts can also extend into the gluteal region.

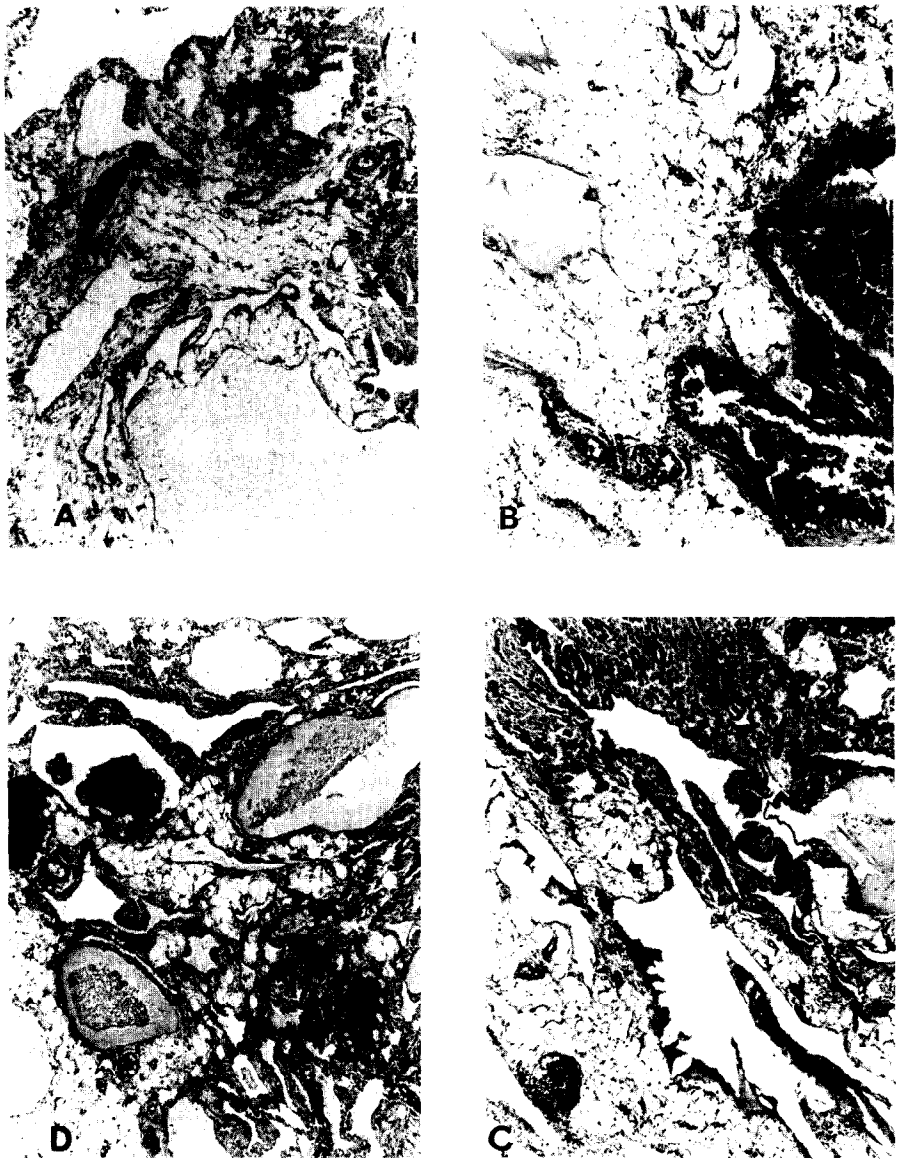

FIGURE 2. *(A to D) Photomicrographs exhibiting multiple lymph channels lined by flat endothelial cells with muscular wall visible in larger channel in center of field with scattered lymphocytes in walls of adjacent tissues.*

These benign neoplasms are congenital in origin, slow growing, and have limited connections to normal lymphatic channels. Ewing[5] believed these tumors occurred in children and adults as multilocular cavernous and cystic tumors, originating along the spinal column and ramifying into the pelvis behind the kidney or colon, upward to the liver and spleen into the omentum. Lymphangiomas have three common characteristics: they are isolated from normal structures; they appear to have arrested development and differentiation; and they exhibit persistent growth of primitive lymphatic systems.

Embryologically there are five regional primitive lymphatic sacs which normally develop into chains of lymph nodes:[6] the paired jugular sacs lateral to the internal jugular veins, an unpaired retroperitoneal sac at the root of the mesentery, and the paired sacs adjacent to the sciatic veins. These sacs form chains of lymph nodes which drain the head, neck, arm, mesentery, hip, back, and leg, respectively. These regional primitive lymph sacs are generally thought to be developmental sites of lymphangiomas. Most authors agree lymphangiomas arise at these sites by continued growth of congenitally misplaced primitive lymphatic tissue which fails to acquire venous connections or as continued endothelial outgrowth of veins. Other explanations include lymph channels which become obstructed secondary to fibrosis, inflammation, trauma, node degeneration, or failure of endothelial secretory function.

In 1877, Wegner[7] histologically divided lymphangiomas into three classifications: (1) lymphangiomas simplex (capillary lymphangioma),

small, thin-walled lymphatic channels not common or found intra-abdominally; (2) cavernous (sometimes malignant),[8] larger thin-walled channels, more common but rare intra-abdominally; (3) cystic (always benign) composed of large cystic spaces lined with flat endothelium, but common retroperitoneally and intra-abdominally.[9] Harrow[10] believed that to be classified as a lymphangioma, these cysts should microscopically contain an endothelial or absent lining, small lymphatic spaces, abundant lymphoid tissue in the wall, and lipid-containing foam cells. Characteristically, these neoplasms are usually incapsulated but have serous or chylous fluid and flat endothelium from increased cyst pressure. Lymphocytes or lymph nodes may be found in the lymphoid tissue as well as nerves, blood vessels, fat, connective tissue, or fibrosis with edema and round cells.[11,12]

Lymphangioma tend to present in the third decade, but no sex predilection has been established. Preoperative diagnosis is usually that of a malignant tumor because of the varied and bizarre symptoms. In Thrupp's[13] series 57.2 per cent had asymptomatic abdominal masses, while 23.8 per cent had infections or hemorrhagic complications, and 19 per cent were postmortem or operative findings. Intestinal obstruction, peritonitis, rupture, or infection may also be presenting symptoms. However, most tumors present with an increasing abdominal or flank mass and a dull flank pain with a "full sensation." Our case presented with a febrile illness and an abdominal mass. Diagnostically, roentgenograms may disclose a soft tissue mass with displacement of an organ or organs. Intravenous urography, arteriography, and gastrointestinal series may confirm this but are nonspecific radiologic signs. Lymphangiography generally is not used because the preoperative diagnosis of a lymphangioma is not usually entertained.

Treatment of such neoplasms consists of preoperative bowel preparations if mesenteric involvement is suspected. Some authors have advocated radiation treatment to the involved area, followed by excision of the tumor. However, we do not believe this is necessary, and a complete en bloc excision of the cystic mass is the treatment of choice, if feasible. Marsupialization or simple drainage is an alternative if complete excision is not feasible. Larson *et al.*[14] recommend sclerosing solutions applied to the cyst lining if the cysts are unroofed or incompletely excised.

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