

Extrarenal retroperitoneal angiomyolipoma: case report and review of the literature

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Abstract

Angiomyolipomas are rare benign tumors that usually occur in the kidneys. Extrarenal angiomyolipomas are extremely uncommon. We describe a case of angiomyolipoma arising in the retroperitoneum that was successfully treated by arterial embolization and surgical excision. The literature on retroperitoneal extrarenal angiomyolipoma and its differential diagnoses is briefly reviewed.

Key words: Angiomyolipoma—Retroperitoneum—Fatty—Tumor—Angiography

Case report

A 35-year-old Chinese woman who was otherwise healthy visited our hospital (Kaohsiung Medical University Hospital) in October 2002 because of gradually increasing abdominal girth and abdominal fullness during the previous year. Physical examination showed a large, smooth, nontender, and immovable mass in the right abdomen. It was palpable from the right costal margin to the right anterior superior iliac spine and extended beyond the midline of the abdomen. The results of laboratory examination, including a differential blood cell count and biochemistry, were within normal limits, except for an elevation of lactate dehydrogenase (647 μL ; normal range, 180–460 μL). Ultrasonography showed a large hyperechoic tumor mass. Computed tomography (CT) disclosed a huge (20 × 9 × 22 cm) retroperitoneal mass displacing the right kidney ventrally. The mass was composed of fatty tissue and hypervascularity (Fig. 1). The abdominal aortogram and selective angiograms of the celiac artery, superior mesenteric artery, right lumbar artery, and right renal artery showed a large hypervascular tumor arising from the right lumbar artery (Fig. 2A) and branches of the right renal artery (Fig. 2B).

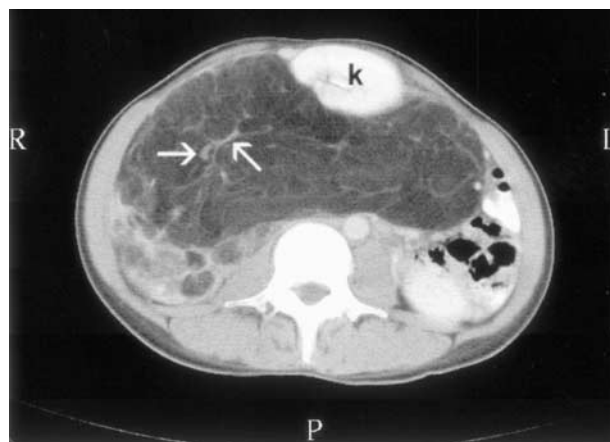


Fig. 1. Contrast-enhanced CT of the abdomen demonstrates a huge retroperitoneal fatty mass filling the entire abdomen. The right kidney (K) is displaced ventrally. Note the rich vascularity of the tumor (arrows).

Because the tumor was hypervascular, we considered transarterial embolization before surgery. The lumbar artery feeding the tumor was successfully embolized with Gelfoam pledget and metallic coils. The right renal artery was difficult to approach.

During surgery, a huge, well-encapsulated retroperitoneal mass was found, which had displaced the right kidney to the left upper quadrant area and occupied the entire right side of abdomen. The tumor was easily dissected from the right kidney and surrounding tissues; the resected tumor weighed 2800 g, with dimensions of 23 × 22.5 × 7.2 cm. The tumor was encapsulated, gray-white, and firm (Fig. 3). Microscopically, it was found to be an angiomyolipoma, composed mostly of mature adipose tissues, numerous blood vessels, and proliferative smooth muscle. Immunohistochemically, the proliferative smooth muscle cells were positive for α -smooth muscle actin (α -superior mesenteric artery), the

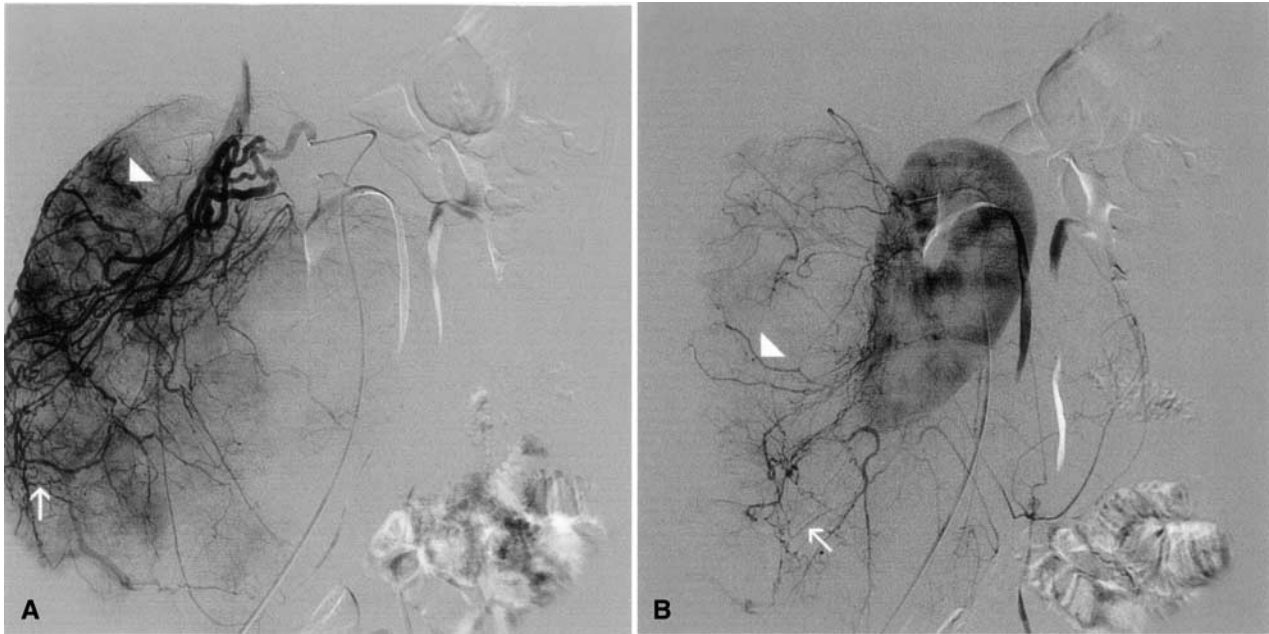


Fig. 2. Abdominal aortogram shows a large hypervascular tumor arising prominently from the (A) right lumbar artery and the (B) right renal artery. The tumor vessels are tortuous (*arrowheads*), enlarged, and resemble a corkscrew (*arrows*).

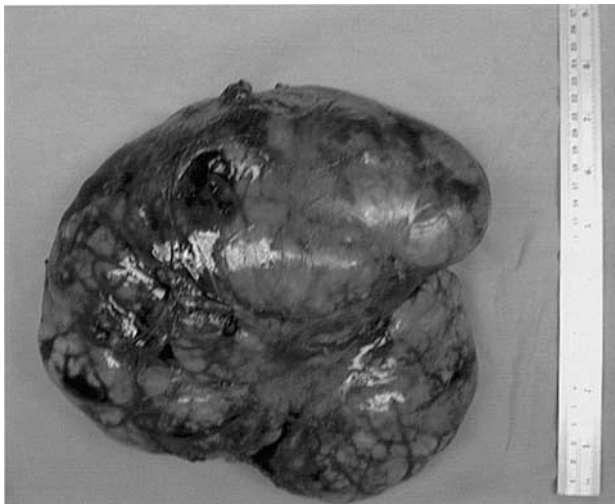


Fig. 3. Gross specimen of the resected tumor. The tumor is well encapsulated, gray-white, and firm. The dimensions are $23 \times 22.5 \times 7.2$ cm.

tumor cells were positive for S-100 protein, and the vascular component was positive for CD34. Recovery was uneventful and the patient was discharged 1 week after surgery.

Discussion

Angiomyolipoma is a rare benign soft tissue tumor that contains smooth muscle, thick-walled vessels, and mature fat and is generally involved in one or both kidneys. Extrarenal angiomyolipomas are very uncommon, and the liver is the

most common extrarenal site. Other sites reported include bone [1], colon [2], heart [3], lung [4], parotid gland [5], skin [6], spermatic cord [7], gynecologic regions [8–10], and the retroperitoneum [11–14]. Extrarenal retroperitoneal angiomyolipoma is extremely rare to our knowledge, with no more than 10 cases having been reported in the literature.

Tuberous sclerosis sometimes occurs in patients with renal angiomyolipoma but has not been reported in patients with extrarenal angiomyolipoma, and the reasons for this are unclear. The patient described in our report had no evidence of tuberous sclerosis. Extrarenal retroperitoneal angiomyolipoma also has been reported to occur with early gastric cancer [12]. In our patient, gastroenteroscopy and colonoscopy were performed, disclosing superficial gastritis and external compression at the ascending and descending colon, respectively.

The radiologic differential diagnosis of retroperitoneal fatty tumors must include liposarcoma, lipoma, leiomyoma with fatty change, and angiomyolipoma. Usually, the correct diagnosis is made after laparotomy, despite numerous preoperative imaging studies. Angiomyolipoma is usually mistaken for liposarcoma. However, it is important to differentiate retroperitoneal angiomyolipoma from liposarcoma because liposarcoma is malignant and should be treated with radical surgery. In contrast, angiomyolipoma is considered cured after tumor excision. Wang et al. [15] reported that analysis of the CT characteristics of linear vascularity, aneurysmal dilatation, bridging vessel, and the beak sign can be helpful in diagnosing retroperitoneal angiomyolipoma, and this report was helpful in making the differential diagnosis in our case.

Like renal angiomyolipoma, extrarenal retroperitoneal angiomyolipoma has a propensity to bleed, and the risk is correlated with the size of the tumor [16]. A huge tumor can be managed with selective arterial embolization to reduce intraoperative bleeding (as in our case) or to stop bleeding during emergencies [17]. In conclusion, selective arterial embolization and simple excision constitute safe and effective management for extrarenal retroperitoneal angiomyolipoma.

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