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Retroperitoneal Angiomyolipoma: Rare Entity

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ABSTRACT

Extrarenal angiomyolipomas (ERAMLs) are rare tumors that present as incidentalomas upon imaging for other conditions. Retroperitoneal ERAMLs present a unique challenge from a diagnostic and treatment standpoint as they can mimic other benign and malignant retroperitoneal tumors.

Keywords: Retroperitoneal, Angiomyolipoma, female.

Introduction

Angiomyolipomas (AMLs) are rare complex mesenchymal neoplasms typically arising within the kidney and are composed of mature adipose tissue, smooth muscle cells, and thick-walled blood vessels. Renal AMLs account for 1% of renal lesions, occurring more commonly in women with an overall incidence in the general population of 0.07–0.3%. Renal AMLs are generally felt to be more like choristomas than hamartomas since kidneys do not normally contain smooth muscle or adipose cells [1].Angiomyolipomas can also occur in other locations (liver, nasal cavity, oral cavity, colon, lung, skin, adrenal glands and bladder)[2].Extrarenal angiomyolipomas (ERAMLs) represent a very rare subset of tumors that often present as incidentalomas upon imaging for other conditions[1]. The retroperitoneum, as a primary site, is the third most common location next to kidney and liver. Although most of the cases are benign, some malignant cases have also been reported; in fact, in many cases, due to the large size of the tumor, the presence of the malignancy is difficult to be excluded and represents an important problem of the differential diagnosis [2].Due to extremely rare occurrence of this tumor, we report the present case.

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Case Report

A 50 year old lady presented in surgical OPD of MBGH, Udaipur with complaints of abdominal fullness, weight gain of 5 kg in last 3-4 months despite decrease in appetite. She also complained of dragging sensation and back pain. There was no history of pressure symptoms related to bladder & rectum. General Examination was unremarkable except mild anaemia with mild pedal edema. On palpation there was firm nontender mass extending from umbilical region into pelvic region of abdomen. There was no hepatosplenomegaly& lymphadenopathy. On Percussion there was dull note.

MRI - T1 weighted MR imaging showed in retroperitoneal region had homogenous signal intensity similar to psoas muscle. T2 weighted showed hyperintense lesion.T1 out phage showed indian ink artifacts. Fat saturation MR imaging showed hypointense mass. Ultrasound (USG) guided needle aspiration of the mass revealed small fragments of fibrofatty tissue comprising of mature adipocytes against the background of fat droplets & hence provisional diagnosis of lipoma was given. The patient underwent exploratory laparotomy; very large vascular soft tumor mass was seen arising from retroperitoneal

region which was excised & sent for histopathological examination. Grossly, a large soft tissue mass (STM) measuring 24×20×10cm and weighing 4.5 kg was received .External Surface was grey white gelatinous at places. On Sectioning, it was partly solid and partly cystic. Solid surface showed large hemorrhagic areas and multiple circumscribed grey white areas. The cysts varied in size from 0.3 to 1 cm in diameter and were filled with mucinous material. Microscopy revealed mature adipose tissue, tortuous thick-walled blood vessels, and bundles of smooth muscle in a fibrofatty & myxoid background.

Discussion

Distinct from renal AMLs, ERAMLs are extremely rare tumors with less than 60 reported cases worldwide in the literature [1]. Friis and Hjortrup reported the first ERAML (1982) in a 22-year-old female presenting with abdominal pain and weight gain who was found on exploratory laparotomy to have an 11 kg retroperitoneal AML[3]. Ditonno et al. [4]have reported the largest series of ERAMLs, involving 40 cases. In their report, the liver was the most common extrarenal location (N = 18), followed by the uterus (N = 7), retroperitoneum (N = 4), and head and vagina (N = 2 each) as well as one each involving the penis, nasal cavity, hard palate, abdominal wall, fallopian tube, spermatic cord, and colon. Other reported uncommon sites include the mediastinum, [5,6] duodenum, appendix, stomach, and adrenal glands[7].

Retroperitoneal ERAMLs present a unique diagnostic challenge since they must be distinguished from other retroperitoneal masses including retroperitoneal sarcomas, atypical lipomas, adrenal adenocarcinomas, leiomyomas with fatty change, and renal cell carcinomas [1]. Although the majority of ERAMLs are benign, 2 cases of metastatic and recurrent ERAMLs have been reported. Gupta et al. described a case of a 29-year-old male with a history of tuberous sclerosis and a retroperitoneal AML which metastasized to the liver and mediastinum 19 years after initial diagnosis and resection. The second case involved an 80-year-old female who developed metastasis to liver and bone one year following surgical resection of a retroperitoneal AML. Although malignant transformation is difficult to predict, high mitotic activity within the primary tumor was a common factor in both metastatic cases[8].

Minja et al. depicted that only 16 cases of retroperitoneal ERAMLs have been reported, making the retroperitoneum the second most common extrarenal location of AMLs [1].Ionica Daniel Vilcea et al. reported a case of retroperitoneal ERAML presented as irreducible inguinal hernia[2].Among patients with retroperitoneal ERAMLs, the average age was 45 years (ranging from 22 to 80 yrs.) with a male: female ratio of 1 to 5.3. Sixty-nine percent of patients with retroperitoneal ERAMLs presented symptomatically with nonspecific abdominal pain, 13% presented with incidentalomas, and another 13% with abdominal fullness.

Retroperitoneal ERAMLs differed widely with respect to size, ranging from 6 cm^3 to 7980 cm^3 and weighing between <1 kg and 11 kgs[1].Additionally, certain ERAMLs variants, most notably the epitheloid variants, are thought to be the most aggressive, suggesting a higher likelihood of metastatic transformation and distant spread[8].

Histologically, angiomyolipomas are described as having three tissue components: convoluted thick walled blood vessels, smooth muscle cells and mature adipose tissue; the proportion of the tissue components is variable, one or the other may prevail, thus three subtypes were described: angiomatous, myomatous, or lipomatous type[2].

The presence of perivascular epithelioid cells (PEC) is often used to characterize angiomyolipomas since these cells show immunoreactivity for muscle markers (epithelial membrane antigen, keratin, vimentin, desmin, and actin) and HMB-45. Positive immunoreactivity for HMB-45, a monoclonal antibody raised against a melanoma-associated antigen, is characteristic of AMLs and can be used to differentiate AMLs from other similar appearing lesions such as liposarcomas, lipomas, leiomyosarcomas or, leiomyomas[1].

Fig 1: Cut Section- was partly solid and partly cystic. Solid surface showed large hemorrhagic areas and multiple circumscribed grey white areas

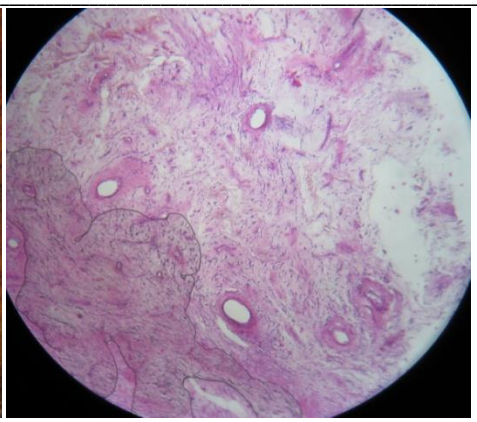

Fig 2: it shows thick & thin walled blood vessels with smooth muscle cells component

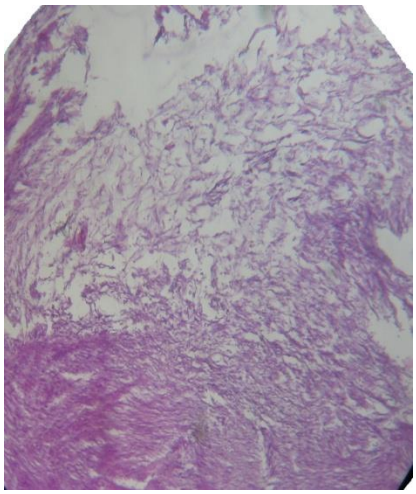

Fig 3: it shows mature adipose tissue & smooth muscle cells

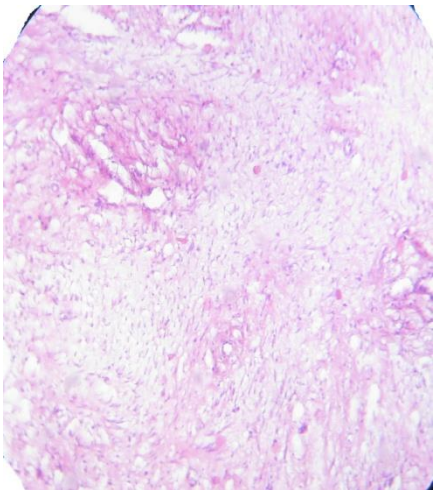

Fig 4: it depicts abundant of mature adipose tissue & smooth muscle cells component. It was partly solid and partly cystic. Solid surface showed large hemorrhagic areas and multiple circumscribed grey white areas. The cysts varied in size from 0.3 to 1 cm in diameter and are filled with mucinous material

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