Case Report

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Retroperitoneal Angiomyolipoma in a Woman of Childbearing Age: A Case Report

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ABSTRACT

Introduction: Extrarenal angiomyolipoma is an uncommon disease entity and is rare in the retroperitoneum. This lesion shows three benign components, including mature adipose tissue, thick-walled blood vessels, and smooth muscles.

Case Report: This report describes a 37-year-old woman who presented with incidental right retroperitoneal mass in prenatal checkup. The patient underwent excisional surgery. Histological examinations and the immunohistochemical study revealed angiomyolipoma. Based on the diagnosis, the patient received no more treatment. After 3 years of close follow-up examinations, no recurrence was observed.

Conclusion: This diagnosis is often confused with many other entities in retroperitoneum. Thus, imaging and histologic correlation are required. Proper diagnosis is essential for further patient management and avoids unnecessary treatment.

Introduction

Angiomyolipomas, previously classified as hamartomas, are now categorized as benign tumors [1]. Histologically, the lesions were most often definitive with the three typical components, including mature adipose tissue, thick-walled blood vessels, and smooth muscle. Tumors are entirely composed of adipose tissue leading to misdiagnosis and mimicking liposarcoma [2]. Angiomyolipoma is the prototype of a heterogeneous family of lesions unified by the presence of HMB45-positive myoid cells, which is mentioned as the perivascular epithelioid cell [3]. Most angiomyolipomas

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are asymptomatic, and they are more common than other angiomyolipomas, affecting 13 per 10,000 adults. They are more found in patients with tuberous sclerosis, where they frequently are accompanied by cysts and rarely by renal cell carcinoma. They are rarely identified before adolescence in patients without tuberous sclerosis. Their incidence is more common in women than men and they sometimes propagate quickly during pregnancy, suggesting that hormones may play a role in stimulating the progression of angiomyolipoma [4].

Angiomyolipomas are typically originated in the kidneys of tuberous sclerosis patients. They are also occasionally seen in patients with polycystic kidney disease, with neurofibromatosis type 1, or with von Hippel-Lindau syndrome. Less frequently, they have been observed in tissues, such as the liver, oral and nasal cavity, heart, large intestine, and lungs [5]. Extrarenal angiomyolipomas are extremely uncommon [6]. Angiomyolipoma is benign but epithelioid variants of angiomyolipoma can relapse and metastasize [7]. In the immunohistochemical study, most of the tumor cells were positive for Human Melanoma Black 45 (HMB-45) (100%) and Smooth Muscle Actin (SMA) (100%) [8]. HMB-45 immunoreactivity is associated with ultrastructural striated organelles that strictly resemble pre-melanosomes; however, no evidence of melanogenesis has been reported in this tumor [9]. It has been well-known that ultrasound and CT scans can properly diagnose angiomyolipoma of the kidney in 86% of cases. Rare cases of extrarenal angiomyolipoma have been described as they are more challenging to diagnose on imaging because they prefer to have no adipose tissue [1].

Angiomyolipomas have variable imaging appearances on multiple imaging modalities and can confuse precise diagnosis in an extrarenal location [10]. However, a preoperative radiological diagnosis of angiomyolipoma is probable. The demonstration of intratumoral fat and central vessels is supportive in the diagnosis [11]. Nevertheless, in our case, as in the others, the correct diagnosis was made only after laparotomy [12]. Current management choices for treatment consist of observation, embolization, and resection. Protocols for treatment are generally based on the patient's symptoms or the size of the lesion [13]. Typically, symptomatic and asymptomatic angiomyolipomas larger than 4 cm in size are generally treated with arterial embolization or resection [14]. However, the indications for treatment are somehow controversial because of the innate risk of spontaneous hemorrhage and consist of bleeding, pain, large tumor size (often mentioned as >4 cm), females of childbearing age, and poor emergency or follow-up care. For instance, numerous angiomyolipomas are small and asymptomatic (over 50% of cases) and are managed by surveillance only [15].

Case Presentation

A 37-year-old healthy woman, who was planning for a pregnancy, was referred to the gynecologic department in Sina hospital affiliated to the Tehran University of Medical Sciences for a prenatal checkup. Routine prenatal lab tests were normal. The patient's family history and past medical history were unremarkable. In physical examination, mild swelling in the right side, below the ribs was identified by the physician and she underwent imaging. In the abdominopelvic ultrasonography, a right side retroperitoneal mass of about 15 cm in the greatest dimension was reported. No other imaging modality was done. Because the mass had a large size, the asymptomatic patient underwent surgery and after excising the mass from the right kidney, the specimen was sent to the pathology department of Sina hospital. She well tolerated the procedure without complications. Received specimen for pathology examination composed of the right kidney with the attached perinephric fat. The external surface of the kidney was smooth and grayish tan. In incision, parenchyma showed a normal appearance. The mentioned mass was 18x13x8cm in size and was attached to the hilum of the right kidney. The external surface of the mass was smooth and dark brown. Cut sections showed an encapsulated tan-yellow surface with multiple hemorrhagic areas. On the microscopic examination, a neoplastic tissue composed of three components was seen: mature adipose tissue, epithelioid cells, and dilated blood vessels. The epithelioid cells have abundant clear to lightly eosinophilic granular cytoplasm with distinct borders and round nuclei. No mitotic activity or necrosis was observed.

Differential diagnoses were an adrenal cortical tumor, liposarcoma, melanoma, renal cell carcinoma, paraganglioma, etc. For a definite diagnosis, an immunohistochemical study was done. Regarding immunohistochemical findings, most epithelioid cells were positive for HMB-45, vimentin, SMA, and Melan-A and negative for CK, CD10, S100, PAX8, myogenin, calretinin, C-Kit, desmin, EMA, synaptophysin, inhibin, CD68, and MDM2. About 1% of tumor cells were positive for Ki67 (proliferative activity index). According to morphologic features and immunohistochemical staining, retroperitoneal angiomyolipoma with epithelioid features was confirmed (Figures 1 and 2). Other differential diagnoses were excluded. Based on the diagnosis, the patient received no additional treatment. After 3 years of close

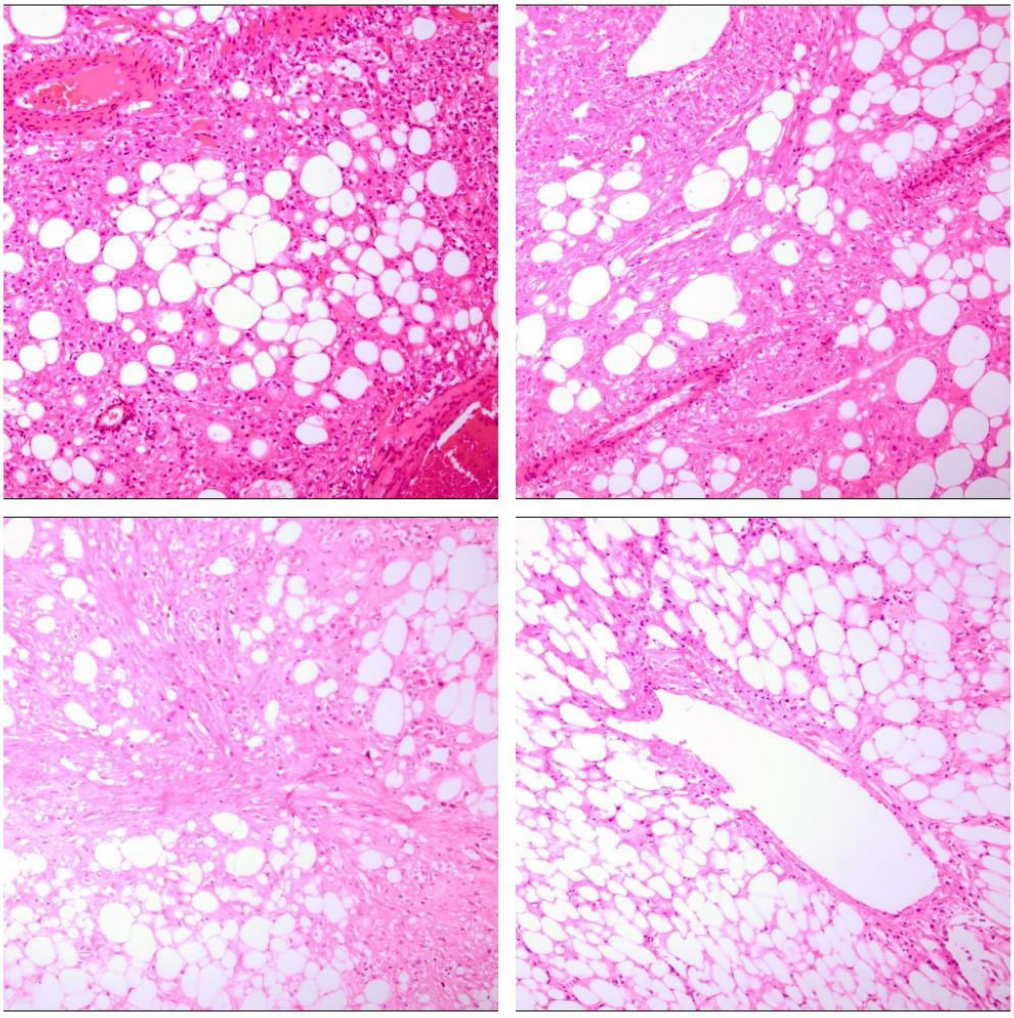

Figure 1. Sections show neoplastic tissue composed of adipose tissue, epithelioid cells, and thick-walled vessels (H&E staining).

follow-up examinations, no recurrence was observed and the patient is still asymptomatic.

Discussion

Extrarenal angiomyolipoma is a rare disease entity and is infrequent in the retroperitoneum [16]. Although both angiomyolipomas and leiomyomas with fatty change presenting as huge retroperitoneal tumors are unusual, and consequently, are not well-known by surgical pathologists, they are benign and must be recognized from liposarcomas [17]. Angiomyolipoma is mostly benign; however, a special subtype (epithelioid angiomyolipoma) may act more aggressively than the typical type. Tuberous sclerosis-related angiomyolipoma is larger, multiple, and more probable to cause spontaneous hemorrhage than the sporadic form. Tumors with hemorrhage are larger [18]. Angiomyolipomas have a higher possibility of rupture through pregnancy [19]. The first case of "Angiomyolipoma" was initially designated by Fischer in 1911 and the term "Angiomyolipoma" was created by Morgan in 1951. Originally, angiomyolipoma was described to be a type of hamartoma or choristoma [20].

Angiomyolipomas are exceptional mesenchymal tumors, established principally from the renal parenchyma, but can also originate in other parts (liver, nasal cavity, oral cavity, colon, lung, skin, adrenal glands, and bladder). Regarding the retroperitoneum, based on the initial location, less than 60 cases have been described until now in medical literature, most of them as single case reports [21]. In another study, renal angiomyolipomas were found in 0.13% of the population with only eleven retroperitoneal angiomyolipomas described in the English literature [22]. However, both an increase in the use of imaging, as well as advances in imaging technology, have caused an increase in the recognized

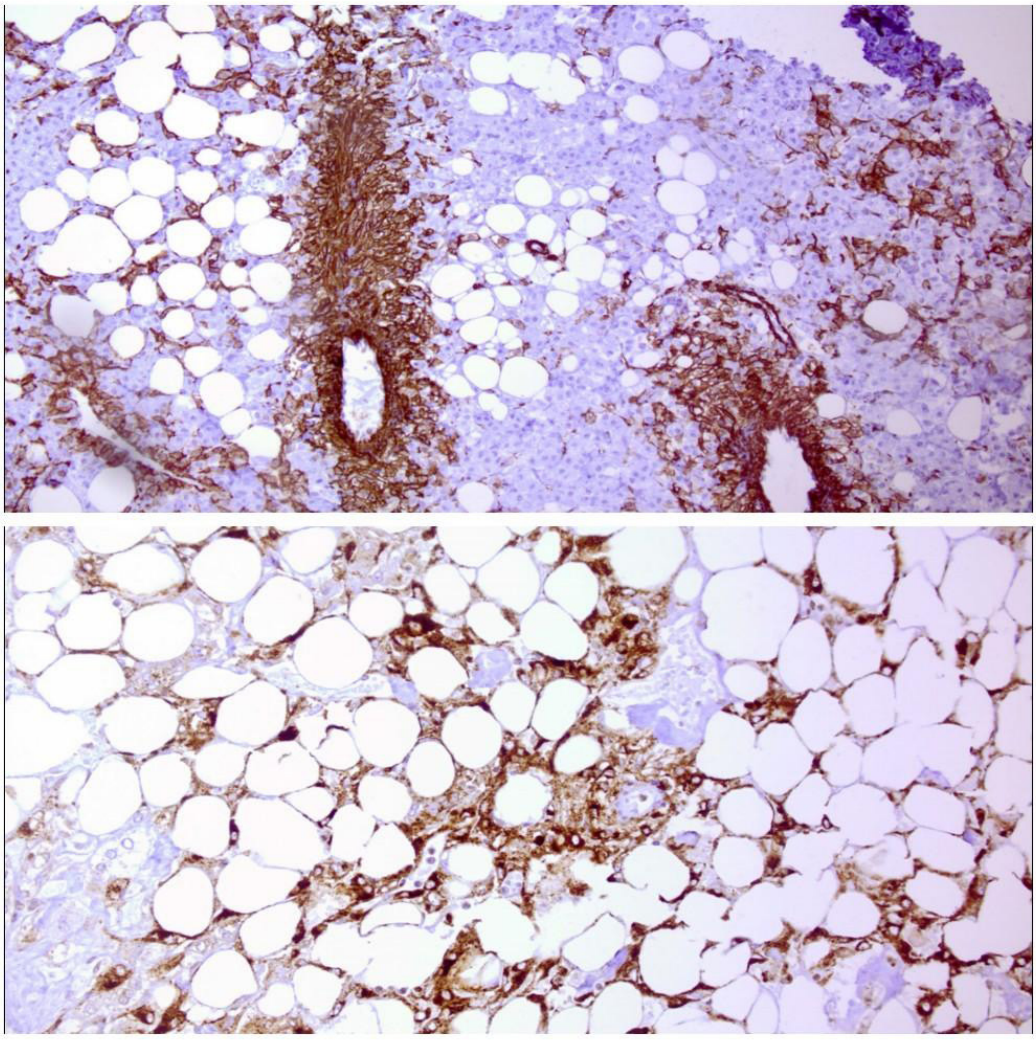

Figure 2. Up: SMA-positive cells in blood vessel walls, Down: HMB-45-positive epitheloid cells.

cases in the general population [23]. Proper diagnosis of angiomyolipoma can avoid unnecessary surgery because conservative medical management with persistent surveillance has been proven to be effective for angiomyolipoma, while surgical resection is the treatment for liposarcoma. Imaging and laboratory follow-up for at least 5 years have been optional in patients subjected to surgical resection of angiomyolipoma [24]. Long-term follow-up resulted in a high angiomyolipoma relapse ratio in patients only with tuberous sclerosis [26].

Conclusion

It is significant that the pathologist recognizes this lesion, even when it is found in the retroperitoneum outside the kidney, and differentiates it from a liposarcoma. Prophylactic intervention for large angiomyolipomas in women of childbearing age should be considered to prevent hemorrhagia in pregnancy.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

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Conflict of interest

The authors declared no conflict of interest.

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