الورم الشحمي العضلي الوعائي في الحوض
تقرير عن كتلة إليوية نادرة

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الملخص

الورم الشحمي العضلي الوعائي (AML) هو ورم حميد، أكثر شيوعًا في الكلى، وأقل شيوعاً في الكبد، ونادراً ما يوجد في الأعضاء الأخرى. والتوضع الحوضي يعد نادر الحدوث، وقد يصل إلى حجم كبير، ويتظاهر بأعراض غير مألوفة. ويصف تقريرنا حالة AML عملاقة في الحوض خلف البريتوان في مريضة عانت من خدر ونمل في الطرف السفلي الأيمن لمدة عام وراجعت بشكوى كتلة إليوية تزداد حجماً مع السعال والوقوف. وتتطلب الاستئصال الكامل فتح البطن بوضعية استخراج الحصاة مع إجراء شق مساعد في الإلية. والدراسة النسيجية أكدت التشخيص. وتعدُّ هذه الحالة الأولى المنشورة من حيث التوضع الحوضي مع تظاهر بشكل كتلة إليوية. ويجب أن تؤخذ هذه الأورام بالحسبان عند مقاربة كتل الحوض.

الكلمات المفتاحية: الورم الشحمي العضلي الوعائي، حميد، حوض، إلية، كتلة.

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ANGIOMYOLIPOMA OF THE PELVIS REPORT OF A RARLY BUTTOCK MASS

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Abstract

Angiomyolipoma (AML) is a benign tumor, most common in the kidney, less in the liver, and rarely found in other organs. Pelvis angiomyolipomas are rare and may reach large size, and found in unusual organs. Our report describes a case of giant pelvic retroperitoneal AML in a female patient had suffered from numbness and tingling in her right lower limb for one year and presented as a buttock mass that becomes larger with cough and standing. Complete excision required laparotomy in the lithotomy position with an assistant incision in the buttock. The histopathological examination confirmed the diagnosis. This is the first published case in terms of pelvic location with a buttock mass presentation. This tumor should be taken in consideration of the pelvic mass approach.

Keywords: Angiomyolipoma, benign, pelvis, buttock, mass.

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Introduction:

Angiomyolipoma (AML) is a benign tumor[1], about 80% of cases are sporadic, and 20% associated with tuberous sclerosis[2-3]. In the past AMLs classified as hamartomas, but now we thought they are PEComas (perivascular epithelioid cell tumors[1]). Commonly exists in the kidney and liver, but it is rare in retroperitoneal and pelvic organs[3]. Patient presents with increasing abdominal girth urinary or digestive symptoms and sometimes spontaneous rupture. We report the first case of giant pelvic retroperitoneal AML that arise as a buttock mass initially misdiagnosed as a perineal hernia or a soft tissue sarcoma.

Case presentation:

A 50-year-old Syrian woman presented with a painless buttock mass caused tingling and numbness for one year. The mass become larger with cough and standing, no history of previous trauma, operations ore medicines. Her menstrual cycle is normal and regular.

Physical examination refers to a mass arises in the ischiopubic area with cough, like hernias. Abdominal examination, biochemistry and hematology results are unremarkable. CEA and CA 125 in the normal rang. Ultrasonography (figure 1) shows a pelvic mass at the right side of the uterus.

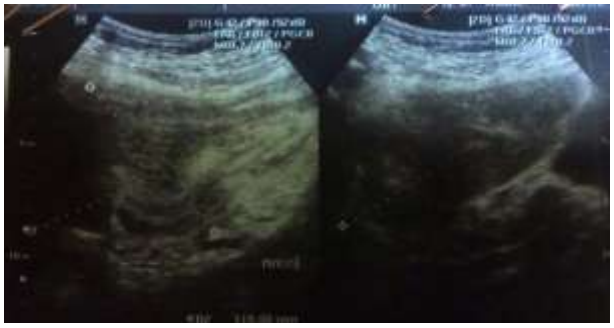

Figure (1): Ultrasonography shows a pelvic mass.

Abdominal computed tomography (CT) with intravenous contrast (figure 2) demonstrated a right retroperitoneal fatty mass situated in the pelvis and extended to the ischiopubic fossa.

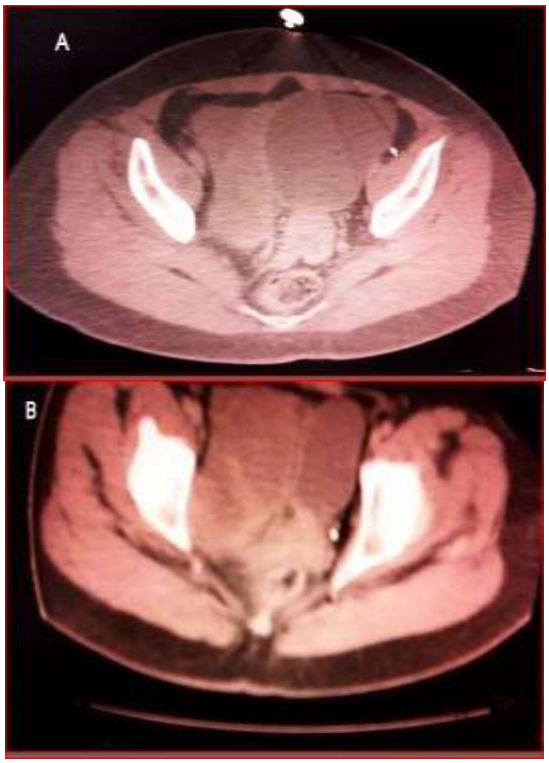

Figure (2): Abdominal computed tomography (CT) with intravenous contract demonstrated a right retroperitoneal fatty mass (A) situated in the pelvis and extended to the ischiopubic fossa (B).

In the lithotomy position laparotomy, we found a giant mass 18 cm, soft in texture, right to the uterus, extending to the ischiopubic fossa passing the greater foramen (figure 3). It was easily dissectible from the uterus and bladder. We released it from the ischial tuberosity assisted by another surgical incision in the buttock. This incision was important to prevent any injury to the vascular structure or the sciatic nerve. The patient discharged after two days without any complication. The histopathological examination of the operative specimen made the diagnosis of AML (figure 4). The patient after 30 months is very well, no symptoms, no recurrence clinically or radiologically.

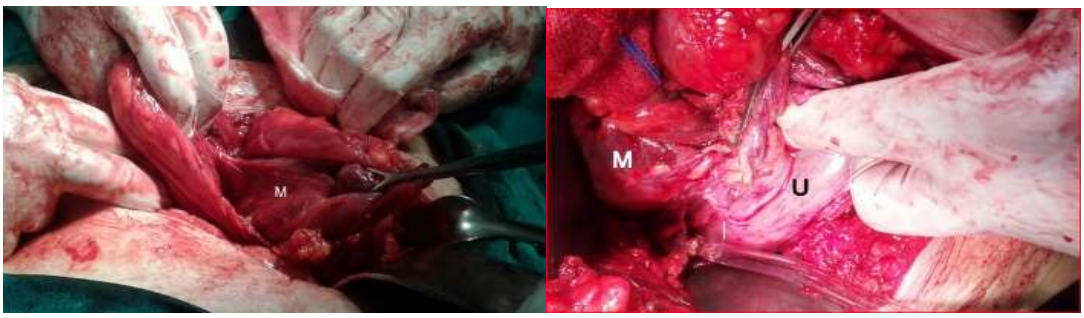

Figure (3): intraoperative image shows a giant mass 18 cm (M), soft in texture, right to the uterus (U)

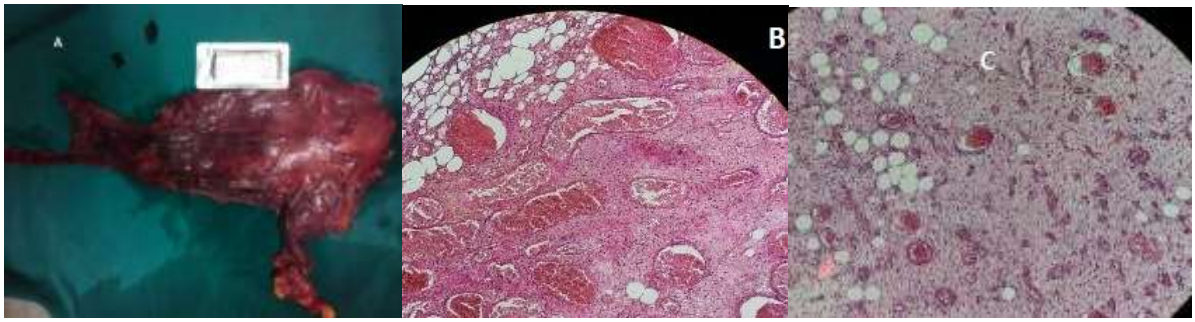

Figure (4): (A) Gross image of the resected mass, demonstrating a well-encapsulated fatty mass, with a smooth outer surface measuring 18 CM. (B, C) Triphasic with myoid spindle cells, islands of mature adipose tissue and dysmorphic thick-walled blood vessels.

Discussion:

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[4].AML is more common in the kidney and the liver than in the soft tissues, occurs extremely rarely in the retroperitoneum[5], pelvic soft tissue[6], spermatic cord[7], and mediastinum[8], scrotum[4], uterus[9],and colon[10].

Extra renal retroperitoneal angiomyolipoma may manifest in a variety of ways including: pain in the abdomen, loin, or back, history of gain in weight, fullness in epigastrium and tiredness. Patients may present with discomfort, malaise, fever, or anorexia. Many cases are asymptomatic, diagnosed on imaging studies performed for other reasons[5]. Sometimes AML presents as acute conditions[11], rupture[12-13], and Aneurysm bleedindg[14]. The studies found AMLs well circumscribed and highly echogenic on ultrasonography because of high fat content[15]. Studies showed these classic findings were observed in only 60% of cases. Adipose tissue demonstrates negative Hounsfield units (HU) on CT scan and is considered the hallmark of AML. Controversy exists, however, regarding the minimum HU value (range, 10 to 100 HU) that confirms this diagnosis[15-16]. Computerized tomography (CT) and computerized tomographic angiography (CTA) are the most commonly used imaging modalities to investigate AMLs. Wang et al.[17] analyzed the radiologic abdominal CTs characteristics of retroperitoneal extra renal AMLs (ERAMLs) in an effort to distinguish them from liposarcomas.

MRI does not appear to have an advantage over CT[14], except when intravenous contrast administration is contraindicated.

A definitive diagnosis of angiomyolipoma is typically made after histopathologic examination of the operative specimen. However, modern imaging modalities, particularly CT scan and MRI scan, have made it possible to identify these lesions in vivo.

AMLS tend to grow slowly, accelerated growth maybe observed in larger masses and multiple tumors. Approximately 77% of tumors that are smaller than 4 cm are asymptomatic, although 82% of AMLs that are larger than 4 cm produce symptoms[11]. Our case describes an 18 cm AML arises in an unusual place and products unusual symptoms as numbness and tingling. Anthony Kodzo[18] reports 30 cases of retroperitoneal AML varies in presentation and treatment, but none of retroperitoneal extending to the buttock. Tsutsumi[19] reported a huge retroperitoneal AML about 3.5 kg. In this case, we describe the buttock presentation of retroperitoneal AML with unremarkable symptoms and radiologic characters. The DDX lead to STS5. THE large size and lack of remarkable symptoms suggest the benign pathology.

Surgery, and embolization in less often tumors, is the primary treatment for ERAMLs[20-21]. Gregory[22] described the using of Radiofrequency ablation of large renal angiomyolipomas.

Surgical excision is indicated for symptomatic, complex appearing, radiologically enlarging, or large ERAMLs, which also have a higher tendency to bleed in patients who present symptomatically with retroperitoneal hemorrhage, selective arterial embolization has been used effectively to control hemorrhagic lesions in hemodynamically unstable patients, resulting in tumor involution and subsequently allowing for elective resection or clinical observation[5-20]. Fearing of rupture, we suggested the surgical treatment. To prevent recurrence, complete en-bloc enucleation was done assisted with another incision. According to this case we have to put the AMLs in the DDX and approach of pelvic and buttock mass.

Conclusion:

Extra renal retroperitoneal angiomyolipoma is a rare tumor which could be confused with other tumors; this case highlights the need to expect this AML in the approach of any buttock or retroperitoneal masses.

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