

A CASE OF ANGIOMYOLIPOMA OF THE TESTIS

حالة ورم عضلي شحمي وعائي في الخصية

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ملخص الحالة

سيتم هنا عرض حالة ورم عضلي شحمي وعائي في الخصية عند رجل عمره 35 سنة عانى من كتلة مؤلمة في الخصية اليسرى منذ مدة أسبوع. راجع المريض قسم الطوارئ حيث أظهر تصوير الخصية بالأشعة فوق الصوتية وجود كتلة متغايرة في الخصية اليسرى مع تموت وتشكل عدة خراجات. تم إجراء استئصال للخصية حيث أظهر الفحص النسيجي للكتلة وجود نسيج شحمي ناضج مع أوعية دموية متعرجة سميكة الجدران وحزم من العضلات الملساء وهو ما يتوافق مع تشخيص الورم العضلي الشحمي الوعائي في الخصية. يعتبر هذا الورم السليم من الحالات النادرة الحدوث وقد أوردت حالات قليلة جداً في الأدب الطبي على مستوى الصنف. يجب الأخذ بالاعتبار عند تدبير هذه الحالة حجم الورم ووجود الأعراض والخصائص المتعلقة بالمريض وخاصة خطر النزف. معظم الأورام العضلية الشحمية الوعائية العرضية تكون بحدود 4 سم والتشخيص النهائي لها يكون عبر الفحص النسيجي.

ABSTRACT

A case of testicular angiomyolipoma is presented herein. A 35-year old male with a history of one week duration of painful left scrotal mass, with previous diagnosis of epididymitis presented to the emergency room with severe left scrotal pain. His scrotal sonography demonstrated that his left testicle was heterogeneous with evidence of left testicular infarction with multiple abscess formation. Left orchidectomy performed and the histopathologic study reported a tumor composed of mature adipose tissue, tortuous thick walled blood vessels and bundles of smooth muscle, corresponding to testicular angiomyolipoma. This benign tumor is rare and there are very few cases reported on at the scrotal level. Its management should take into account the size of the tumor, the presence of symptoms and factors characteristic of the patient, particularly the

risk for hemorrhage; the majority of symptomatic angiomyolipoma measure around 4 cm. Histopathologic study regularly provides the definitive diagnosis.

INTRODUCTION

The majority of tumors of the testis is derived from germ cells and account for 90-95%. Non-germ testicular tumors constitute just 5% of all testicular neoplasms mostly comprised of malignant lymphoma, Leydig cell tumors and Sertoli cell tumors.¹ Angiomyolipoma (AML) is a common benign mesenchymal tumor that is usually found in the kidney.² AML in tissues other than the kidney is rare and can affect the visceral organs including the kidney, lung,³ liver,⁴ nasal cavity⁵ small and large intestine,⁶ prostate and uterus, as well as the retroperitoneum,^{7,8} pelvis,⁹ pancreas,¹⁰ skin,^{11,12} testis,¹³ and scrotum^{14,15,16,17} and to our knowledge, very few

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cases have been described in the testis to date. Here we report a case of testicular AML in 32-year old man with detailed findings.

Angiomyolipoma is a rare tumor (0.13% to 0.3% in the general population) that mainly involves the kidneys, presents in women over 50 years of age, and is particularly associated with tuberous sclerosis. Originally described by Fischer in 1911, Morgan gave it its present name in 1951. (AML) are mesenchymal tumors of the kidney consisting of varying proportions of vascular, immature smooth muscle and mature fat cells. AML are only rarely malignant, with only 12 cases of malignant transformation reported in the literature, and the vast majority are benign and do not necessitate treatment unless they are very large and at risk of bleeding.

They occur most commonly in the kidney and less occasionally in the liver, bone, retroperitoneum, pelvis and soft tissue. Though most AML occur sporadically, they can also be associated with tuberous sclerosis an autosomal dominant condition characterized by seizures, mental retardation, skin lesions (ash leaf spots) and hamartomas of other areas of the body. Less commonly AML can be associated with von Hippel Lindau, von Recklinghausen, or autosomal dominant polycystic kidney disease.

CASE PRESENTATION

A 35-year old male with a history of one week duration of painful left scrotal mass, with previous diagnosis of epididymitis presented to the emergency room with severe left scrotal pain. His scrotal sonography demonstrated that his left testicle was heterogeneous mainly hypoechoic inechopattern without evidence of blood flow and evidence of left testicular infarction with multiple abscess formation. The patient underwent scrotal exploration which ended by left orchidectomy. His pathologic examination of the mass revealed a tumor composed of mature adipose tissue, tortuous thick walled blood vessels and bundles of smooth muscle. The tumor involving most of the testis with remnant foci of epididymis also the tumor was involving the spermatic cord. Areas of abscess formation with pus,

hemorrhage and granulation tissue formation were seen. Immunohistochemistry of the lesion revealed smooth muscle actin (SMA+) positively highlighted the smooth muscle bundles, and endothelial marker CD34 positively highlighted the vascular spaces, features corresponding with angiomyolipoma. His tumor markers Beta HCG, alphafeto protein, were within normal value, and his abdominal CT scan revealed small Rt renal cortical cyst in lower pole, multiple small sub cm paraaortic L.N were noted with intact pelvi- abdominal solid organs.

DISCUSSION

Testicular AML is not officially listed in the World Health Organization histological typing of male testicular tumors. Therefore, the diagnosis and treatment strategy for testicular AML has not been established. Angiomyolipoma is an uncommon, benign tumor that is rarely located in the scrotum. The first case of AML in the testis has been described by Lane et al in 2004,¹⁵ and reported a case of AML diagnosed by microscopic findings without immunohistochemical analysis. The tumor in the present case revealed a tumor composed of mature adipose tissue, tortuous thick walled blood vessels and bundles of smooth muscle, the tumor involving most of the testis with remnant foci of epididymis also tumor involving the spermatic cord, in addition to the superimposed inflammatory process as the presence of areas of abscess formation with pus, hemorrhage and granulation tissue formation. This atypical presentation of this case as emergency case of acute scrotumwith abscess, led us to make the diagnosis by pathologic examination and immunohistochemistry of the lesion which revealed smooth muscle actin (SMA) and endothelial marker CD34 positively highlighted the vascular spaces. In conclusion, pathological and immunohistochemical study strongly indicated benign testicular AML with a non-germ cell origin. Although his abdomino-pelvic CT revealed para-aortic small nodulesit was considered a manifestation of the multicentric nature of AML or benign swelling due to the previous inflammatory changes.

CONCLUSIONS

Angiomyolipoma is an uncommon, benign tumor that

is rarely located in the scrotum. Its management should take into account the size of the tumor the presence of symptoms and factors characteristic of the patient, particularly the risk for hemorrhage; the majority of symptomatic angiomyolipoma measure around 4 cm. Histopathologic study regularly provides the definitive diagnosis.

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