

A case of angiomyolipoma of the spermatic cord and testicle

Roberto Giulianelli, Luca Albanesi, Francesco Attisani, Stefano Brunori, Barbara Cristina Gentile, Luca Mavilla, Gabriella Mirabile, Francesco Pisanti, Giorgio Vincenti, Teuta Shestani, Manlio Schettini

Villa Tiberia Clinic, Urology Department, Rome, Italy

Summary

Angiomyolipomas (AML) are mesenchymal tumors of the kidney consisting of varying proportions of vascular, immature smooth muscle and mature fat cells.

A rare case of testicular AML is described. A 53 year old male with a history of congenital motor defects, mental retardation, and hypertension, presented to the emergency room with sudden onset, severe left testicular pain. Scrotal sonography demonstrated an hypoechoic mass in the patient's left testicle.

The patient was offered and underwent a trans-inguinal exploration of the left testicle which ended in a left inguinal orchiectomy. Pathologic examination of the mass revealed medium to large calibre thick-walled blood vessels with ectatic lumina, surround by sclerotic, fibrous smooth muscle bundles in a fatty milieu. Immunohistochemistry of the lesion demonstrated positive staining for smooth muscle actin (SMA+) and endothelial marker CD34. The lesion did not, however, stain positively for smooth muscle antigen S100 or melanocytic antigen HMB-45.

KEY WORDS: Angiomyolipoma; Testicular tumor; Orchiectomy.

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INTRODUCTION

Angiomyolipomas (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 (1). They occur most commonly in the kidney and less occasionally in the liver, bone, retroperitoneum, pelvis and soft tissue. The incidence of AML in the general population is believed to be between 0.3% and 7% and AML, though in modern series, they account for only 1% of surgically extirpated tumors.

Though most AML occur sporadically, they can also be associated with tuberous sclerosis (TS), a 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* (VHL), *von Recklinghausen*, or autosomal dominant polycystic kidney disease (citations). In this case report, we describe a rare case of testicular AML treated at our institution.

CLINIC CASE

A 53 year old male with a history of congenital motor defects, mental retardation, and hypertension, presented to the emergency room with sudden onset, severe left testicular pain. His physical exam was significant for an indurated and painful left testicle and epididymis. His blood tests were non-contributory with the exception of slight leukocytosis to 12.65 h/mm³.

A scrotal sonogram was performed which showed bilateral testicular microcalcifications as well as an enlarged and hypoechoic left epididymal head. He had normal arterial and venous flow bilaterally.

The patient was prescribed antibiotics and anti-inflammatory therapy. After two weeks, his symptoms had not resolved and a repeat scrotal ultrasound was performed. His repeat scrotal sonogram demonstrated that his right testicle was normal in size, shape and echotexture with the exception of multiple 1-2 mm microcalcifications. His left testicle was 38 x 30 x 30 mm, contained multiple microcalcifications, and was hypoechoic. There was dilation of the left pampiniform plexus.

Because of the hypoechoic mass in the patient's left testicle, the patient was offered and underwent a transinguinal exploration of the left testicle which ended in a left inguinal orchiectomy. Gross examination of the testicle revealed a well circumscribed, 4 x 3 x 3 cm, soft brown mass replacing most of the testicular parenchyma. Within the mass there was widespread hemorrhage.

Pathologic examination of the mass revealed medium to large calibre thick-walled blood vessels with ectatic lumina, surround by sclerotic, fibrous smooth muscle bundles in a fatty milieu. The thin rim of testicular tissue remaining around the tumor displayed diffuse hemorrhage and necrosis within the seminiferous tubules and interstitial xanthogranulomatous flogistic infiltrate with blood cell extravasation and siderophages. The tunica albuginea was irregularly thickened. The epididymis appeared to be uninvolved.

Immunohistochemistry of the lesion demonstrated positive staining for smooth muscle actin (SMA+) and endothelial marker CD34. The lesion did not, however, stain positively for smooth muscle antigen S100 or melanocytic antigen HMB-45.

DISCUSSION

Angiomyolipomas are rare lesions which occur predominantly in the kidney. While the overall incidence is estimated to be 0.3%-7% in the general population, the incidence in patients with TS is approximately 50%. AMLs exhibit a female predominance in patients both with and without TS; however in patients with TS, AMLs tend to be diagnosed during the 20's and 30's and are often small, asymptomatic and multifocal (2). Sporadic cases of AML are usually diagnosed later (in the 5th to 8th decade) and are generally single, unilateral and larger. Because of their size, they are more likely to be symptomatic.

Though there are 12 case reports in the literature of malignant AML, the vast majority are benign and do not necessitate treatment unless they are very large and at risk of bleeding. Because of this and because these tumors can often be reliably diagnosed with CT scan, only 1% of surgically extirpated renal lesions are AMLs. Though AMLs were once believed to be hamartomatous lesions, the rare incidence of malignant transformation and the presence of clonal mutations suggests that they are true neoplasms (1). Though AMLs are classically comprised of three tissue types, some AMLs are predominantly only one or two of these tissue types. These tumors can be mistaken for other lesions: leiomyomatous AML can be mistaken for leiomyomas and excessively fatty AML can be mistaken for liposarcomas. The vast majority of our knowledge regarding AMLs comes from renal AMLs. Several markers are used to diagnose them, but smooth muscle actin (SMA) is positive in almost 100% of these lesions while melanocytic marker HMB-45 is positive in 80-100% of these lesions. Other markers frequently used are CD34, an endothelial marker, and S100, a smooth muscle marker (1). Extra-renal AMLs occur much more infrequently. To our knowledge there is only one other case of testicular AML reported in

the literature (6), though there is also a case involving the spermatic cord (3) and spleen (4). Given the rarity of these tumors, there is no consensus regarding the imaging, diagnosis, or treatment of these lesions. As in Saito's report (6), our patient's tumor was clearly not of germ cell origin. It was SMA+ but did not stain for HMB-45. A series of cutaneous HMB-45 negative AMLs has also been reported, prompting the author to propose that these tumors should be called angioleiomyomas with fat instead of angiomyolipomas (5). Despite the histopathologic nomenclature, until further studies are done to establish reliable methods of diagnosis and treatment, we feel that it is most prudent to perform a transinguinal orchiectomy on these tumors. While this may seem aggressive, in both reported cases, these lesions have replaced the majority of the testicle by the time they are detected. Since the amount of viable testicular parenchyma left was minimal and both reported patients have been symptomatic, surgical extirpation appears to confer little disadvantage.

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Correspondence

Roberto Giulianelli, MD
Villa Tiberia Clinic - Rome, Italy
roberto.giulianelli@virgilio.it

Luca Albanesi, MD
Francesco Attisani, MD
Stefano Brunori, MD
Barbara Cristina Gentile, MD
Luca Mavilla, MD
Gabriella Mirabile, MD
Francesco Pisanti, MD
Giorgio Vincenti, MD
Teuta Shestani, MD
Manlio Schettini, MD
Villa Tiberia Clinic - Rome, Italy