

· Letters to the Editor ·

Scrotal aggressive angiomyxoma mimicking inguinal hernia

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Dear Sir,

I am Dr Yao-Chou Tsai, from Department of Urology, Buddhist Tzu Chi General Hospital, Taipei Branch, Taipei, Taiwan, China. I write to you to present a case report of scrotal aggressive angiomyxoma (AAM).

AAM is a rare, locally infiltrative mesenchymal benign neoplasm with a predilection for involving the female pelvis, perineum, vagina and cervix. Its occurrence in men is much less frequent and only scattered case reports have been reported. To our knowledge, only 43 cases of AAM occurring in men have been reported in the English-language literature [1]. AAM in the male inguinal area can present as an inguinal mass that is usually diagnosed and treated as inguinal hernia [2, 3]. Herein, we report an unusual case of a 40-year-old man with an AAM involving the scrotum, who underwent a herniorrhaphy at a prior hospital. The AAM was not removed. To our knowledge, this is the first documented case in which scrotal AAM was missed during surgical exploration for inguinal hernia.

A 40-year-old man complained of a right scrotal mass, which had grown slowly over the previous 10 years. No local pain or urinary tract symptoms were noted. The scrotal mass was initially diagnosed as an inguinal hernia and a herniorrhaphy was performed at a prior hospital.

However, the scrotal mass still remained *in situ* after the operation. Therefore, he came to our hospital for further management.

On physical examination, the right scrotal mass was elastic, non-tender, non-transilluminated and not reducible. Scrotal ultrasonography revealed a homogeneous, hypochoic, extratesticular, extraepididymal, well-demarcated mass lesion. No calcification was present. Pelvic computed tomography (CT) demonstrated a 5.0 × 5.1 × 5.4 cm heterogeneous mass in the right scrotum with scrotal wall thickening. The testicle was displaced cephaladly. Chest X-ray radiograph revealed no significant abnormal finding. Laboratory studies were normal for blood count, urine analysis, renal function, liver function, serum alpha-fetoprotein, beta-human chorionic gonadotropin and lactic dehydrogenase. The clinical impression was a paratesticular tumor and a wide excision was planned.

In surgery, an incision through a previous herniorrhaphy wound was done and a large tumor was found within the spermatic fascia (Figure 1). The boundary of the tumor was well circumscribed and away from the right testicle and epididymis (Figure 2). The cut surface of the tumor was yellowish and gelatinous, and no necrosis or cystic changes were present. The frozen section of the tumor showed a loose myxoid and fibrous background containing blood vessels of variable caliber.

The evenly distributed stromal cells were wavy, spindle or stellate in shape and had delicate cytoplasmic processes. Mitoses and cellular pleomorphism were lacking (Figure 3). The findings are consistent with AAM. The tumor was excised completely and the patient was discharged on the first postoperative day. The patient has remained free of local recurrence for 12 months since the operation.



Figure 1. The tumor was well enclosed with right testis by a fascia continued from spermatic cord.

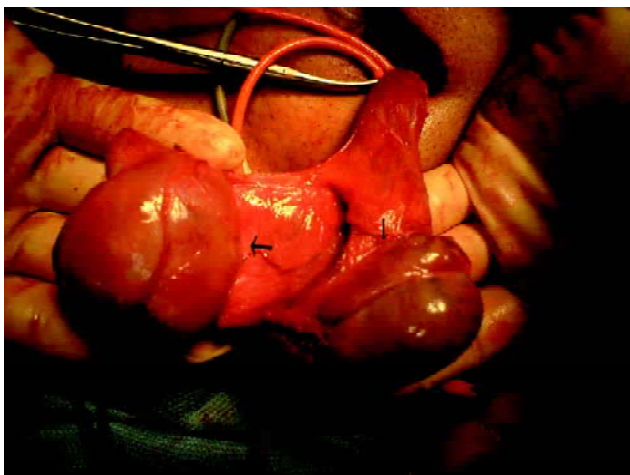


Figure 2. Gross findings during operation. Thick arrow indicated AAM that was separated from right testicle and epididymis (arrow).

Since Steeper and Rosai [4] first described nine cases of aggressive angiomyxoma in female adults in 1983, its incidence has increased year by year. Male AAM tends to be an asymptomatic round mass lesion that is sometimes initially diagnosed as an inguinal hernia. Previously reported AAM that have been found incidentally during herniorrhaphy have been resected smoothly with satisfactory postoperative results [2, 3]. However, in clinical practice, it is possible that a scrotal AAM is missed if a concurrent inguinal hernia exists. To avoid this, detailed preoperative physical examination and confirmative tumor removal is necessary.

Preoperative imaging studies do not always provide a correct diagnosis because the tumor usually shows an invasive and hypervascular picture that makes the tumor seem malignant [5]. Gray-scale ultrasonography demonstrates a hypoechoic, homogeneous and well-demarcated mass without significant flow using color Doppler ultrasonography. On CT, AAM is hypodense relative to muscle and hypoattenuating with specific swirling internal architecture on enhanced scans. The tumor has high signal intensity using T2-weighted magnetic resonance imaging. The swirling internal architecture of lower intensity after intravenous contrast enhancement is also seen within the high signal tumor.

Grossly, AAM is yellowish in color and usually well demarcated and encapsulated. The cut surface demonstrates gelatinous and glistening material with delicate, white, fibrous strands. Microscopically, AAM demon-

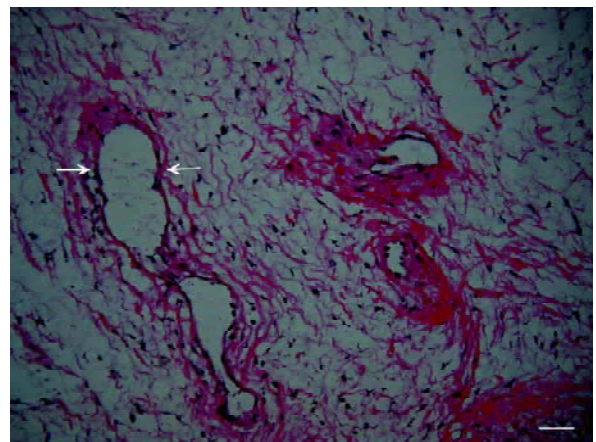


Figure 3. Microscopically, various sized vessels (arrow) with spindled or stellate-shaped tumor cells were scattered in the loose myxoid stroma. Scale bar = 50 μ m (H&E, \times 200).

strates stellate and spindle-shaped cells in a loose myxoid stroma without mitosis or nuclear atypia. Vascularization is abundant, and small to large thick-walled vessels together with collagen fibrils are scattered in the hypocellular background.

Aggressive angiomyxoma should be distinguished from other benign and low local recurrent potential lesions such as intramuscular myxoma, myxoid neurofibroma, myxoid or spindle cell lipoma, superficial angiomyxoma and angiomyofibroblastoma. The malignant tumors with widespread metastasis potential should also be considered a differential diagnosis (the myxoid variants of liposarcoma, malignant fibrous histiocystoma and embryonal/botryoid rhabdomyosarcoma) [6].

Characteristically, AAM is locally aggressive and infiltrative without metastasizing potential, with the exception of two cases of death reported by Blandamura *et al.* [7] and Siassi *et al.* [8]. Its local recurrence rate is approximately 37% and average time of recurrence is 9 months to 14 years postoperatively [2, 4].

In conclusion, AAM should be considered when an usually soft tissue mass is found during physical examination or surgical correction for inguinal hernia. Because of its highly recurrent characteristics, wide excision of the tumor and strict postoperative follow up is recommended.

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