Massive recurring angiomyxoma of the scrotum in a obese man

Francesco Rocco,[1] Gabriele Cozzi,[1] Matteo Giulio Spinelli,[1] Bernardo Maria Rocco,[1] Giancarlo Albo,[1] Elisabetta Finkelberg,[1] Isabella Oliva,[1] Paolo Fontanella,[1] Daniela Varisco,[1] Laura Moneghini,[2] Michele Talso,[1] Carlotta Palumbo[1]

[1]Department of Urology , Università degli Studi di Milano, Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Milan; [2]U.O. Anatomia Patologica, Azienda Ospedaliera San Paolo, Milan, Italy

Abstract

Aggressive angiomyxoma (AAM) is a rare mesenchymal benign myxoid tumor, characterized by locally infiltrative nature and high recurrence rate. AAM occurs almost exclusively in adult females, arising from the soft tissue of the pelvic region: to our knowledge, only 43 cases occurring in men have been reported. We report a case of massive recurrence of scrotal AAM in a 46-year-old obese man, who already underwent surgery for the same disease in 2004 and 2005. The mass had a circumference of 106 cm and weighted 30 kg. It was impossible to appreciate the testes and to find the penis. The patient underwent scrotal resection, bilateral orchidopexia and transposition of the penis, by means of a preputial flap. Residual scrotal skin was modeled in order to create a neoscrotum, where the testes were placed and secured with interrupted sutures. Histologic examination showed diffuse angiomyxoma-like lipomatosis. After three months, the patient presented with local relapse which also involved the external urethral orifice.

Introduction

Aggressive angiomyxoma (AAM) is a rare mesenchymal benign myxoid tumor first described in 1983 by Steeper and Rosai.[1] This tumor, characterized by locally infiltrative nature and high recurrence rate (36-72%),[2] was given its name due to the pathological findings of stellate and spindled cells along with variable-sized blood vessels intricately entwined within a myxoid matrix.[3] AAM occurs almost exclusively in adult females, arising from the soft tissue of the pelvic region, perineum, vulva and buttock.[4] To our knowledge, only 43 cases occurring in men have been reported in the literature.[5] We report a case of massive recurring AAM of the scrotum in a 46-year-old obese man.

Case Report

A 46-year-old obese man (237 kg - body mass index 76) presented with a recurring angiomyxoma of the scrotum: he underwent surgery for the same disease in 2004 and 2005. The patient also suffered from hypogonadism, hyperinsulinism and hypercorticosurrenalism. Blood count, renal function, acute-phase proteins, and blood pressure values were normal.

Physical examination evidenced massive enlargement of the scrotum, whose longitudinal diameter was 32 cm, transverse diameter was 34 cm, and circumference was 106 cm (Figure 1). The mass weighted 30 kg. Skin was discromic, with some decubitus ulcers. It was impossible to appreciate the testes and to find the penis. After cardiologic and anesthesiologic assessment, the patient underwent scrotal resection, bilateral orchidopexia and transposition of the penis. Before the surgery the positioning of a vesical catheter by means of a rigid cystoscope was attempted, but the urethral meatus was not found. The scrotum has been incised after positioning some landmark sutures, in order to preserve some tissue for the reconstructive phase. The incision began from the right inguinal region with monopolar electrocautery and Ligasure (TM). The right spermatic cord and testis, which were in the deeper part of the scrotal mass, were isolated (Figure 2). The right emiscrotal mass, weighting about 25 kg, was then resected. The penis was isolated together with a preputial flap with paramedian incision, and a Foley-Tiemann 16 Ch vesical catheter was placed.

A circular incision was performed over the pubic symphysis, and the penis was transposed suturing the cavernous bodies to the skin (Figure 3). Residual scrotal skin was modeled in order to create a neoscrotum, where the testes were placed and secured with 3 interrupted sutures. Histologic examination showed adipose tissue which was loosely arranged with increased interspaces between fat cells, without inflammation. Thin- and thick-walled hyaline vascular channels crossed the adipose tissue like vessels of an angiomyxoma (Figure 4). Widely scattered spindle-shaped cells with ill-defined cytoplasm composed same areas of the lesion. The chronic nature of the lesion was represented by areas with an increased number of dilated vessels and miofibroblastic spindle cells with collagen deposition. The diagnosis was the same of 2004 and 2005.

The postoperative course was prolonged because of the late active mobilization of the patient due to an intraoperative compression of the left deep peroneal nerve, diagnosed by electromyography and treated with anti-inflammatory drugs and physical therapy. After three months, the patient presented with local relapse: the mass, mainly affecting the left hemiscrotum, measured 15 cm of transverse diameter, and involved the penis conditioning a stricture of the external urethral orifice. Now the patient is waiting for a further surgical resection; in the meantime, he has been offered adjuvant therapy with gonadotropin releasing hormone (GnRH) agonist (leuprolide acetate, 3.75 mg/month) and weekly urethral dilations.

Discussion

AAM is a rare mesenchymal benign myxoid tumor, poorly circumscribed and characterized by locally infiltrative nature[1] and relatively rapid growth.[3] AAM occurs almost exclusively in adult females, and, to our knowledge, only 43 cases occurring in men have been reported in the literature.[5] In men, AAM usually involves the scrotum (38%), spermatic cord (33%), perineal region (13%), and intrapelvic organs such as the bladder (8%).[4] Although AAM has always been described as only locally infiltrative, in 1999 Siassi *et al.*[6] described a case of an AAM of the pelvis that metastasized to the mediastinum and lungs in a 63-year-old woman. Macroscopically, AAM is a large, grossly gelatinous, poorly circumscribed[7] and locally infiltrative tumor.[4] Its microscopic features are myxoid stroma, low cellularity and vessels of varying size, sometimes with hyaline thick walls.[7] Cellularity is usually low, and mitoses

Correspondence: Gabriele Cozzi, Clinica Urologica I, Università degli Studi di Milano. Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Milan, Italy.
Tel. +39.3339433071.
E-mail: cozzi.gabriele@gmail.com

Key words: angiomyxoma, scrotum, recurrence, obesity.

Received for publication: 14 May 2011.
Revision received: 8 July 2011.
Accepted for publication: 13 July 2011.

Rare Tumors 2011; 3:e31
doi:10.4081/rt.2011.e31

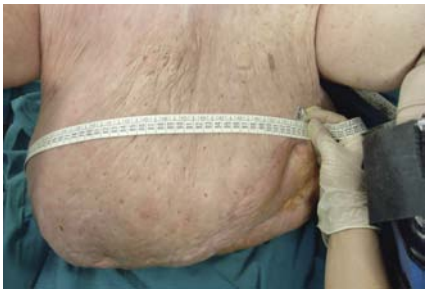

Figure 1. The scrotal mass. The circumference was 106 cm.

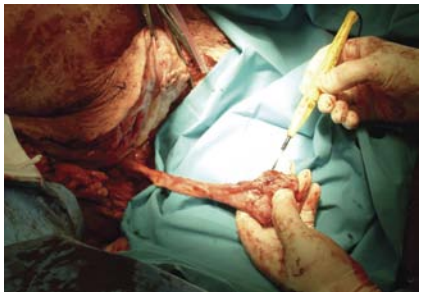

Figure 2. Isolation of the right spermatic cord and testis.

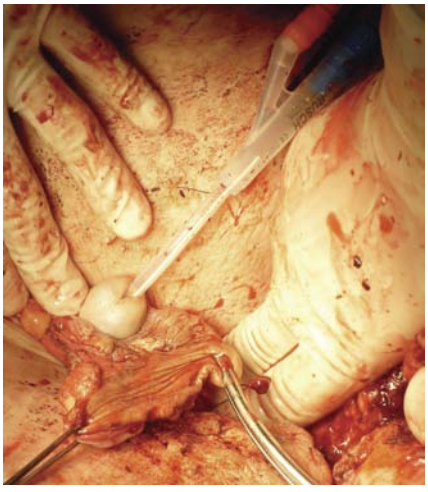

Figure 3. Transposition of the penis through the circular incision over the pubic symphysis.

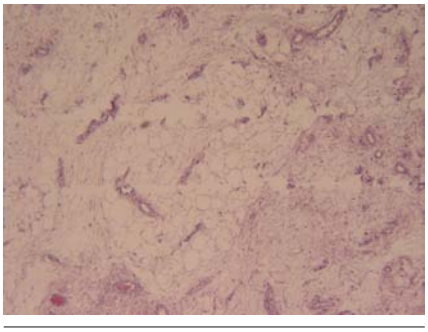

Figure 4. Adipose tissue with edema which scattered fat cells without inflammation. Angiomyxoma-like vessels are present (10X; Hematoxilin/Eosin).

are not seen.[6] The anomalous muscular artery is very specific for diagnosis of AAM, and is the major histological difference form angiomyofibroblastoma.[4] High proliferating cell nuclear antigen (PCNA) immunoexpression, together with the lack of expression of the p21 protein are associated with a high number of recurrences: in fact, when p21 is expressed, it binds PCNA, allowing DNA-repair but no DNA-replicative processes.[7]

AAM often shows an invasive and hypervascular pattern like a malignant tumor, thus preoperative diagnosis by imaging studies is not always correct.[2] Ultrasonography (US) shows a hypoechoic mass, with thin echogenic septa, corresponding to the fibrous strands seen at gross examination.[8] US-guided needle biopsy has been shown to give inconclusive diagnosis.[4] Magnetic resonance imaging (MRI) identifies AAM as an isointense tumor relative to muscle on T1-weighted and hyperintense T2-weighted images.[5] Anyway, most cases of AAM are visualized by computed tomography (CT) scans, which are more accessible to the patients.[4] Both contrast enhanced CT and T2-weighted MRI show a characteristic swirling internal pattern.[5] AAM should be distinguished from other benign tumors affecting the pelvis and the genital tract, such as intramuscular myxoma, myxoid neurofibroma, myxoid or spindle cell lipoma, superficial angiomyxoma, angiomyofibroblastoma and angiomyolipoma; in the differential diagnosis one should also consider some malignant tumors with myxoid stroma, such as myxoid liposarcoma, myxoid malignant fibrous histiocytoma, and embryonal rhabdomyosarcoma.[7] Surgery is the mainstay of treatment in patients with AAM. This tumor tends to infiltrate, thus incomplete extirpation may result in a local recurrence. Local recurrence rate is between 36% and 72%, with multiple recurrences as in this case. Recurrent disease is usually controlled by repeated operations.[2] Radiation therapy combined with the sensitizer razoxane has been described as able to control a recurrent AAM for an unknown time.[9] It has recently been demonstrated that AAM possesses estrogen and/or progesterone receptors, suggesting that this tumor is a hormonally responsive neoplasm and that local recurrences can be treated or prevented with GnRH agonists such as leuprolide acetate, 3.75 mg/month. However, the duration of the response and the optimal treatment schedule are still unknown.[10]

References

1. Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum: report of nine cases of distinctive type of gynaecologic soft-tissue neoplasm. Am J Surg Pathol 1983;7:463-75.
2. Chihara Y, Fujimoto K, Takada S, et al. Aggressive angiomyxoma in the scrotum expressing androgen and progesterone receptors. Int J Urol 2003;10:672-5.
3. Andres LA, Mishra MJ. Angiomyxoma diagnosed in a man presenting for abdominal lipectomy. Can J Plast Surg 2007;15:163-4.
4. Morag R, Fridman E, Mor Y. Aggressive angiomyxoma of the scrotum mimicking huge hydrocele: case report and literature review. Case Report Med 2009;2009: 157624.
5. Idrees MT, Hoch BL, Wang BY, Unger PD. Aggressive angiomyxoma of male genital region. Report of 4 cases with immunohistochemical evaluation including hormone receptor status. Ann Diagn Pathol 2006;10: 197-204.
6. Siassi RM, Papadopoulos T, Matzel KE. Metastasizing aggressive angiomyxoma. N Engl J Med 1999;341:1772.
7. Carlinfante G, De Marco L, Mori M, et al. Aggressive angiomyxoma of the spermatic cord. Two unusual cases occurring in childhood. Pathol Res Pract 2001;197:139-44.
8. De la Ossa M, Castellano-Sanchez A, Alvarez E, Smoak W, Robinson MJ. Sonographic appearance of aggressive angiomyxoma of the scrotum. J Clin Ultrasound 2001;29:476-8.
9. Rhomberg W, Jasarevic Z, Alton R, et al. Aggressive angiomyxoma: irradiation for recurrent disease. Strahlenther Onkol 2000;176:324-6.
10. Shinohara N, Nonomura K, Ishikawa S, et al. Medical management of recurrent aggressive angiomyxoma with gonadotropin-releasing hormone agonist. Int J Urol 2004;11:432-5.