Case Report

Paratesticular aggressive angiomyxoma: A rare case

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Abstract

Aggressive angiomyxoma (AAM) particularly testicular origin is a rare benign mesenchymal myxoid tumor which is locally aggressive, blatant for local recurrence, and may metastasize. It occurs mostly in females of childbearing age and extremely rare in males. AMM particular testicular origin is not reported in literature yet. This is a 65-year-old man who had a right scrotal swelling. Ultrasound scrotum showed a soft tissue tumor of the right testis. The patient underwent radical right orchidectomy of which histopathologically confirmed to be a paratesticular AAM with clear resection margins. There were no signs of local recurrence or metastasis 2 years postsurgical resection.

Key Words: Aggressive angiomyxoma, myxoid tumors, myxoma, soft-tissue tumor, testicular tumor

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INTRODUCTION

Aggressive angiomyxoma (AAM) particularly from testicular origin is a rare benign mesenchymal myxoid tumor which is locally aggressive, blatant for local recurrence, and may metastasize. It occurs mostly in females of childbearing age and extremely rare in males. We report a very rare case of paratesticular AAM which presented as a scrotal swelling.

CASE REPORT

A 65-year-old man presented with the right scrotal swelling for 6 months and enlarging in size. Physical examination revealed a nontender right scrotal swelling measuring 16 cm × 10 cm, soft in consistency. Transillumination examination was positive. Ultrasonography (USG) examination demonstrated a well-defined hyperechoeic mass seen within the right scrotum measuring 3.4 cm × 3.8 cm × 4.8 cm, surrounded by complex locules of fluids, and thickened surrounding soft tissue [Figure 1]. The normal right testis was not visualized. A radical right orchidectomy was done. Intraoperatively, a large right testicular mass measuring 11 cm × 8 cm × 5 cm was found.

Serial sections revealed an ill-defined glistening gelatinous lesion at the paratesticular region measuring 8 cm in largest dimension with adjacent atrophic testis at the periphery. Histologically, it appeared poorly circumscribed with infiltrative border [Figure 2a]. The lesion was hypocellular, composed of uniform and bland-looking spindle to stellate shaped neoplastic cells embedded within the loose myxoid stroma. Numerous small- and medium-sized thick walled vessels are also seen [Figure 2b and c]. No nuclear atypia or mitosis is found. The adjacent atrophic testis was noted at the

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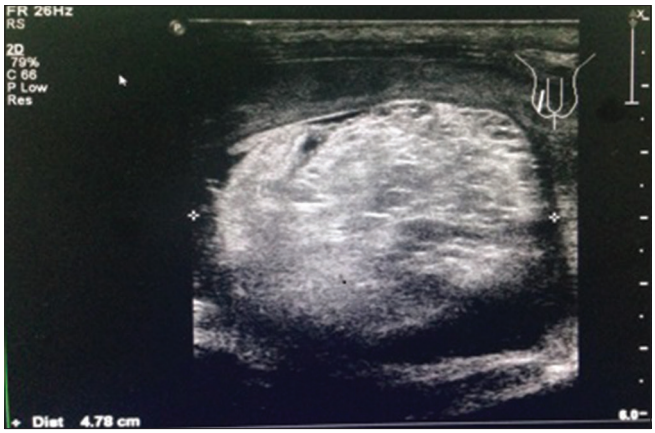

Figure 1: A well-defined hyperechoic mass seen within the right scrotum measuring 3.4 cm × 3.8 cm × 4.8 cm, surrounded by complex locules of fluids and thickened surrounding soft tissue

periphery [Figure 2d]. Immunohistochemically, the neoplastic cells showed diffuse smooth muscle actin (SMA) and desmin immunoreactivity [Figure 2e] and were focally immunopositive for estrogen receptor [Figure 2f]. Progesterone receptor, however, was negative. CD34 and S100 were also negative.

The patient was followed up regularly with USG and computed tomography (CT) scan. He did not have local recurrence or distant metastasis 2 years postsurgical resection.

DISCUSSION

Angiomyxomas are distinctive soft tissue tumors associated with a high risk of local recurrence but the lack of metastatic potential. These tumors occur nearly exclusively in the soft tissues of the pelvis and perineum of adult women. Three types have been identified: AAM, angiomyofibroblastoma, and superficial angiomyxoma.[1] Steeper and Rosai first described AAM in 1983.[2] It is termed aggressive due to its locally infiltrative nature and the high risk of local recurrence up to 72% with multiple recurrences in some cases.[3,4] Local recurrence is considered to be the result of incomplete extirpation and should be controlled by repeated operations.[3] Surgery remains the mainstay of treatment to date. Other treatment modalities such as radiotherapy and hormonal manipulation using tamoxifen, raloxifene, and gonadotropin-releasing hormone analogs were reported.[4,5] Hormonal therapy enhances the feasibility of complete excision in large tumors. In spite of benign tumor, metastasis AAMs were reported.[6] Long-term follow-up with either USG or CT scan is recommended due to its local aggressiveness.[7]

In men, AAM is extremely rare and usually derived from the pelviperineal interstitial tissue involving the scrotum (38%), spermatic cord (33%), perineal region (13%), and intrapelvic

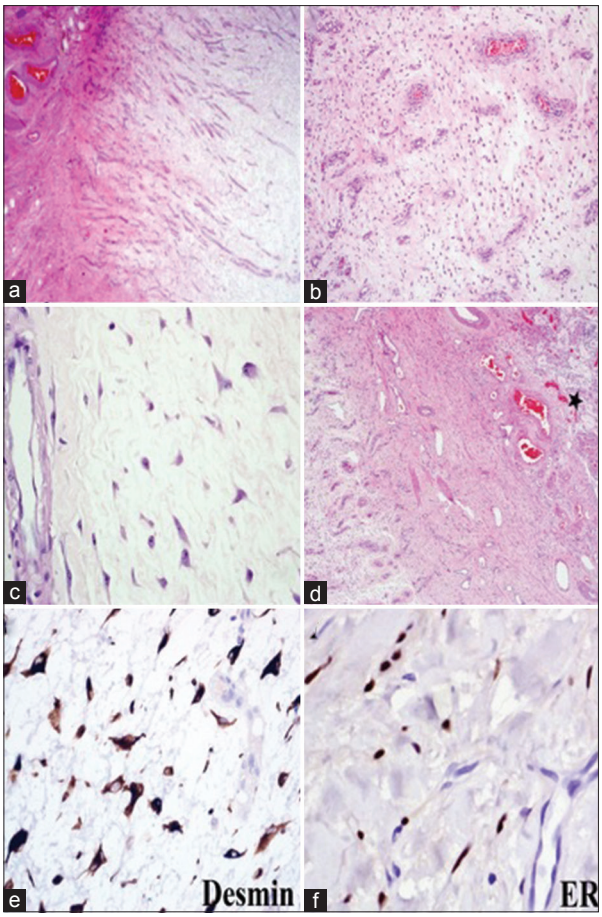

Figure 2: Histological features of aggressive angiomyxoma. (a) Ill-defined lesion with infiltrative border (H and E, ×40). (b and c) The lesion is sparsely cellular, composed of bland-looking neoplastic spindle to stellate cells with small- and medium-sized blood vessels in the myxoid background (H and E, ×100, ×400). (d) Adjacent atrophic testis (asterisk) is also noted (H and E ×40). The neoplastic cells show immunoreactivity for (e) desmin (desmin ×400) and (f) estrogen receptor (estrogen receptor, ×400)

organs (8%).[3,8] AAM in the scrotal region may present as a scrotal mass, often wrongly diagnosed as a hernia, hydrocele, spermatocele, or testicular neoplasm as in the current case. Detailed radiological workup such as USG, CT scans, and magnetic resonance imaging may be helpful in the diagnosis, but histological examination of the excisional specimen is the gold standard for establishing the diagnosis.[9]

We encountered the first case of paratesticular AAM presented as a scrotal mass after reviewing the final histological examination. Macroscopically, AAM typically presented as large (more than 5 cm), grossly gelatinous, and locally infiltrative tumor.[10] Histologically, the tumor appears poorly circumscribed with infiltrative border and consists of uniform and bland-looking spindle to stellate shaped neoplastic cells arranged in a loose myxoid background. Numerous small- and medium-sized thick walled vessels are usually present.[10] Immunohistochemically, the neoplastic spindle cells

are typically immunoreactive for SMA, desmin, and vimentin. Estrogen and progesterone receptors maybe positive in some cases.[10] Classically, the tumor cells are immunonegative for S100 protein and CD34.[2]

Histological differential diagnosis includes angiomyofibroblastoma, cellular angiofibroma, and myxoid variant of solitary fibrous tumor. Unlike AAM, angiomyofibroblastoma is usually well circumscribed with alternating hyper- and hypo-cellular edematous zone with irregularly distributed capillary-sized lacking hyalinized vessels. On the other hand, cellular angiofibroma is typically cellular alternating with wispy collagen bundles. Thick-walled hyalinized vessels are typically seen. Immunonegativity for CD34 has ruled out myxoid variant of solitary fibrous tumor in the current case.

CONCLUSION

Paratesticular AAM is a very rare benign neoplasm which is locally aggressive, blatant for local recurrence, and may metastasize. Surgery is the mainstay of treatment, and subsequent long-term radiological follow-up is recommended.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Vella R, Calleri D. Superficial angiomyxoma of the epidiymis. Presentation of a new case and clinical considerations. Minerva Urol Nefrol 2000;52:77-9.
2. Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum. Report of nine cases of a distinctive type of gynecologic soft-tissue neoplasm. Am J Surg Pathol 1983;7:463-75.
3. Chihara Y, Fujimoto K, Takada S, Hirayama A, Cho M, Yoshida K, *et al.* Aggressive angiomyxoma in the scrotum expressing androgen and progesterone receptors. Int J Urol 2003;10:672-5.
4. Rhomberg W, Jasarevic Z, Alton R, Kompatscher P, Beer G, Breitfellner G. Aggressive angiomyxoma: Irradiation for recurrent disease. Strahlenther Onkol 2000;176:324-6.
5. McCluggage WG, Jamieson T, Dobbs SP, Grey A. Aggressive angiomyxoma of the vulva: Dramatic response to gonadotropin-releasing hormone agonist therapy. Gynecol Oncol 2006;100:623-5.
6. Siassi RM, Papadopoulos T, Matzel KE. Metastasizing aggressive angiomyxoma. N Engl J Med 1999;341:1772.
7. Poirier M, Fraser R, Meterissian S. Unusual abdominal and pelvic tumors: Case 1. Aggressive angiomyxoma of the pelvis: Response to luteinizing hormone-releasing hormone agonist. J Clin Oncol 2003;21:3535-6.
8. Kondo T. Aggressive angiomyxoma in the inguinal region: A case report. J Med Case Rep 2010;4:396.
9. Rehman S, Muqim RU, Gul T, Wazir MZ, Zarin M. Aggressive angiomyxoma of scrotum presenting as an inguinal hernia. Pak J Med Sci 2010;26:478-81.
10. Morag R, Fridman E, Mor Y. Aggressive angiomyxoma of the scrotum mimicking huge hydrocele: Case report and literature review. Case Rep Med 2009;2009:157624.