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Case report

Angiomyofibroblastoma of the spermatic cord: a case report

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Key words

Angiomyofibroblastoma • Male genital tract • Paratesticular • Immunohistochemistry

Summary

Angiomyofibroblastoma (AMF) is a rare benign mesenchymal tumor with tendency to arise in the lower genital tract of middle-aged women, predominately in the vulva. A few cases of AMF in males have been reported involving the scrotum, perineum or spermatic cord. We report a new case of AMF arising in the right inguinal region of a 27-year-old man. The tumor was well-circumscribed, myxoid and measured 30 mm in maximum dimension. On microscopic examination, the tumor was composed of spindle cells without atypia and with less than one mitosis figure per 10 high-power fields. Multinucleated cells and mast cells were observed. The stroma was myxoid and edematous with abundant capillary-sized blood vessels. Immunohistochemical staining showed a strong immunoreactivity for desmin and smooth muscle actin. The tumor cells were negative for estrogen receptors and focally positive for progesterone receptors with a low proliferative index of Ki67 (< 5%). This unusual neoplasm should be distinguished from aggressive angiomyxoma and other myxoid malignant tumors.

Introduction

Angiomyofibroblastoma (AMF) is a rare benign mesenchymal tumor occurring mainly in the vulvovaginal, cervical and inguinal region in women. A few cases of AMF in males have been reported involving the inguinal region [1]. We present a new case of AMF of the spermatic cord.

Case report

A 27- year- old man presented with right inguinal hernia. The patient underwent a surgery, a tumoral nodule of the spermatic cord was found associated to the hernia. The testis and the epididymis were not involved. Surgical excision was performed. Macroscopically, it was a 3 cm, myxoid and well-circumscribed nodule. Microscopic examination revealed a fusocellular proliferation encapsulated by a thick fibrous capsule with alternating hypercellular and hypocellular edematous areas (Fig. 1). The cells were spindle shaped with eosinophilic cytoplasm and elongated nuclei without atypia and with less than one mitosis figure per 10 high-power fields. Cell borders were often indistinct. Multinucleated cells and mast cells were observed within the myxoid and edematous stroma (Fig. 2) intermingled with abundant capillary-sized blood vessels. Immunohistochemical stains showed a strong immunoreactivity for desmin and smooth muscle actin (Fig. 3). The tumor cells were negative for estrogen receptors and focally positive for progesterone receptors with a low proliferative index of Ki67 (< 5%).

Discussion

AMF is a rare tumor first described in the vulva by Fletcher et al. in 1992 [2]. To date 26 cases of male AMF have been reported [1]. Male genital tract AMF involves scrotum, perineum or spermatic cord [3 4]. Grossly, it is mostly well circumscribed, round, ovoid, nodular mass sometimes gelatinous with a soft to rubbery consistency. The cut surface has a grayish-brown homogeneous appearance with no hemorrhage or necrosis. Most cases of AMF measure less than 5cm, and previously reported cases ranged from 0.5 cm to 13.0 cm in greatest diameter [2 5]. On microscopic examination, AMF is well de-

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Fig. 1. H. Ex 100: Alternating hypercellular and hypocellular edematous areas, Myxoid stroma with abundant thick-walled blood vessels.

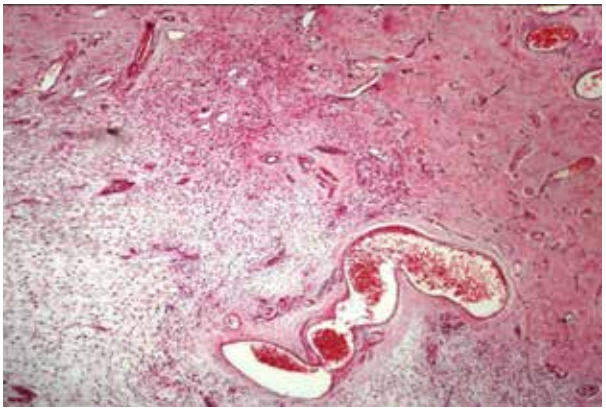


Fig. 2. H.E x200: Multinucleated cells within the mesenchymal proliferation.

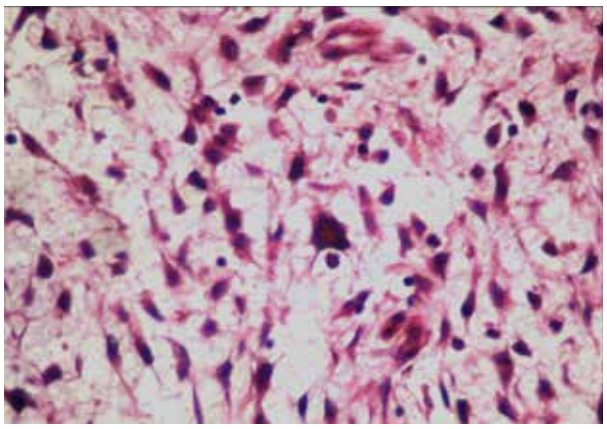
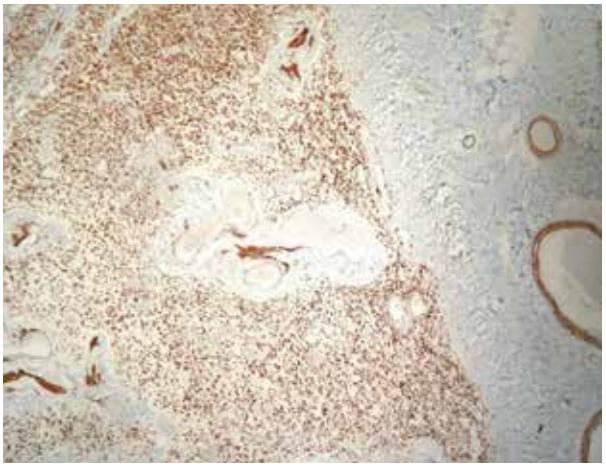


Fig. 3. Positive staining of tumoral cells for smooth muscle actin.



marcated tumor surrounded by a thin fibrous pseudo capsule and presents alternating areas of hypercellularity and hypocellularity with stromal cells that are characteristically aggregated around numerous vessels [1]. The neoplastic cells were spindle-shaped, plasmacytoid, or epithelioid with presence of a variable binucleated or multinucleated cells and clusters of mature adipocytes. The immunohistochemical profile of AMF is variable. Stromal cells show intense and diffuse positivity for vimentin and desmin but they are inconsistently positive for specific muscle actin, smooth muscle actin and CD34. The tumor cells show variable expression of estrogen and progesterone receptors and show a very low Ki-67 index [1 3]. In our case, the immunostaining was negative for estrogen receptors and focally positive for progesterone receptors. The cell origin of AMF remains unclear. Some investigators consider AMF to be a myofibroblastic origin [6].

The most important differential diagnosis of AMF is aggressive angiomyxoma (AAM), which is locally invasive with a high risk of local recurrence and occurs predominately in the pelvic-perineal region. AAM differs from AMF in its infiltrative growth pattern, lower cellularity and a less conspicuous vascular component. It has been suggested that AMF and AAM are related neoplasms, both included in a wide spectrum of angiomyxoid tumors, which exhibit some overlapping features and various combinations of myofibroblastic, fibroblastic and lipomatous differentiation [6]. The differential diagnosis of AMF also includes smooth muscle tumors, peripheral nerve sheath tumors, glomus tumor, chondroid syringoma, myxoid malignant fibrous histiocytoma, angiomyolipoma, spindle cell lipoma and myxoid liposarcoma [7]. It has been proposed that AMF might arise from perivascular stem cells, which are capable of differenciating into fatty and myofibroblastic differenciation [8].

This tumor has no local recurrence or metastatic potential [6].

Local excision with clear margins appears sufficient for the surgical management of AMF [3].

In conclusion, AMF is a rare benign tumor which occurs rarely in male and must be known by the urologists and pathologists. It should be distinguished from aggressive angiomyxoma and other myxoid malignant tumors. The treatment of choice for AMF is simple total excision. There are almost no incidences of recurrences or metastasis after complete excision.

References

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