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Sonographic appearance of angiomyofibroblastoma-like tumor of the scrotum

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Abstract

A variety of extratesticular tumors and tumor-like conditions can occur in the scrotum. Angiomyofibroblastoma is a rare, slow-growing, well-circumscribed mesenchymal tumor, found predominantly in women, and is even a more unusual occurrence in the male genital tract. We describe a case of a 64-year-old male presenting with a scrotal painless mass. On sonography, a well-circumscribed extratesticular mass was present. As no specific imaging findings were present, it was a surprise to learn that the subsequently excised mass proved to be an angiomyofibroblastoma-like tumor. To the best of our knowledge, the sonographic appearance of this tumor in the scrotum has been reported previously in only one case.

Introduction

Angiomyofibroblastoma (AMF) is a rare, slow-growing and usually asymptomatic benign mesenchymal tumor that predominantly arises in the genital tract, vulva, perineum and pelvis in women, while angiomyofibroblastoma-like (AMF-like) tumors develop in the inguinal region, scrotum and perineum in men[(1)]. They usually arise in the 5th–6th decade of life. This type of tumor is derived from perivascular stem cells and has capacity of lipoid and myofibroblastic differentiation. AMF-like tumors have been reported in small series of patients and in few case reports[(2)], the majority being in the inguinal region, but the condition has received little attention in the imaging literature.

We present an unusual case of AMF-like tumor of the scrotum. To the best of our knowledge, the sonographic appearance of this tumor in the scrotum has been reported previously in only one case[(1)].

Case report

A 64-year-old man became aware of a painless, scrotal mass that was slowly enlarging, and was referred for sonography for further evaluation. The patient had no significant medical history and denied having fever or trauma to the scrotum. Physical examination revealed a 3.0 cm painless, slightly mobile and well-circumscribed, rubbery mass adjacent to but separate from the left testis. The skin overlying the mass was normal. Sonography of the scrotum demonstrated normal left and right testes and normal blood flow on Doppler imaging. An additional well-defined solid, oval mass measuring 3.0 × 2.0 cm with similar echogenicity and similar blood flow to the adjacent testes (Fig. 1 and Fig. 2) was demonstrated. This mass was located in the left hemiscrotum but was separate from the testis and epididymis. No hydrocele was identified and both epididymides were normal. This appearance gave rise to a differential diagnosis of a supernumerary testis or a mesenchymal tumor.

The excised specimen, accessed via an inguinal incision, consisted of an oval encapsulated mass measuring 3.5 × 2.0 × 2.0 cm. On section, the center was composed of brown-yellow mucoid tissue. Microscopic sections of the specimen showed the presence of a well-circumscribed, non-cystic mass surrounded by a condensation of fibro-collagen forming a pseudo-capsule. The mass showed variably sized endothelial vascular channels

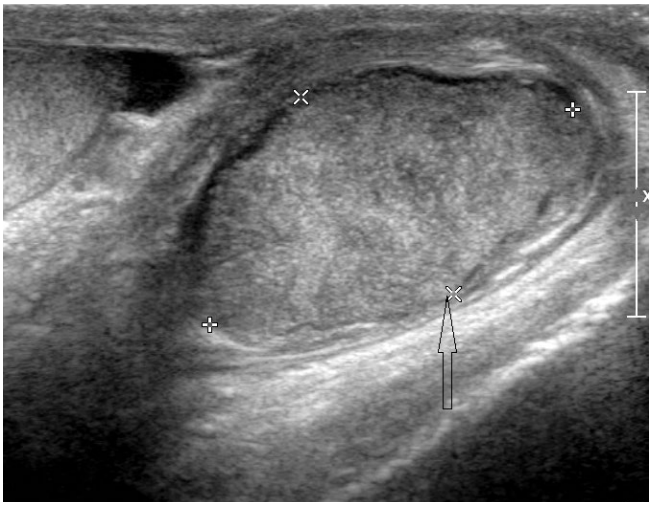

Fig. 1. *Longitudinal scan of the left hemiscrotum demonstrates an oval mass (arrow) with echogenicity similar to that of the adjacent testis*

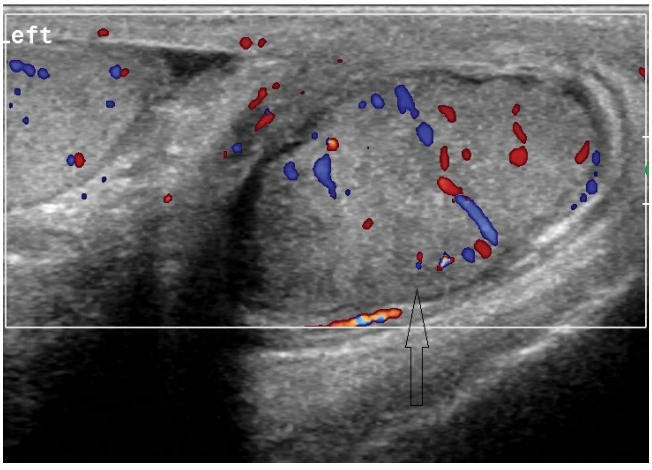

Fig. 2. *Color Doppler scan shows similar blood flow in the mass (arrow) and adjacent testis*

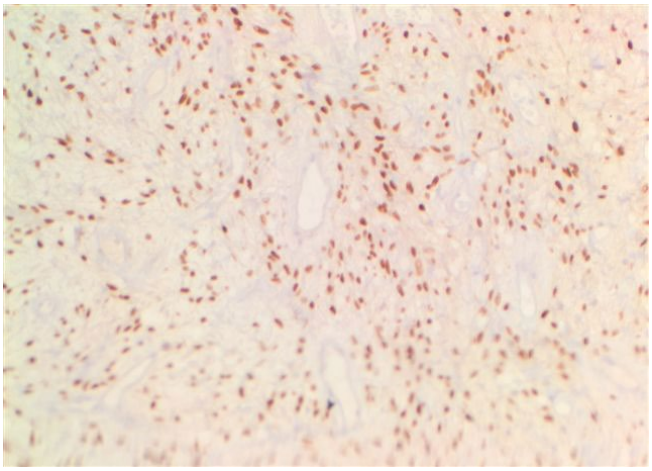

Fig. 3. *Immunostaining – estrogen × 100 showing nuclear positivity in bland fibroblastic stromal cells with negatively stained vascular channels*

lined with bland endothelial cells. Between the vascular channels, there was spindle cell proliferation with alternating hypocellular and hypercellular areas. No mitoses were found in these spindle cells. The spindle cells were strongly positive for CD34 and most of the tumor cells' nuclei were typically and diagnostically positive for estrogen and progesterone (Fig. 3). The SMA and S100 proteins were also positive. The findings were consistent with a para-testicular AMF-like tumor. The patient had an uneventful recovery from surgery.

Discussion

In 1992, Fletcher *et al.*[3] were the first to describe a new type of benign mesenchymal tumor, named AMF, occurring in the female genital tract. The same tumor in the male genitalia, initially named cellular angiofibroma, was renamed *AMF-like tumor* in a report of 11 cases by Laskin *et al*. in 1998[4] due to its similarities with the AMF tumor described in women. These tumors were found to be superficial and well-circumscribed, ranging in size from 2.5 to 14.0 cm[5]. Generally, AMF-like tumors exhibit a benign clinical course, rarely invasive, and usually occur in the 5th–6th decade of life. The most common sites of involvement are the inguinal region, followed by the scrotum[6]. When occurring in the scrotum, they are extratesticular and non-epididymal.

On histology, the tumor demonstrates high vascularity with perivascular fibrinoid hyalinization. Tumor cells are spindle cells, with almost no mitotic activity or atypia. Immunohistochemical analysis shows that the cells are usually positive for CD-34 and desmin, with strong reactions to vimentin, muscle-specific actin, progesterone receptors, and estrogen receptors[5,7].

The recommended treatment for AMF-like tumors is wide surgical resection. Recurrence has been described in only one case, and this occurred 13 years after surgery[4]. It is important to distinguish AMF-like tumors from aggressive angiomyxomas, which are also found in the inguinoscrotal area but have a high risk of recurrence and a malignant locally infiltrative behavior. AMFs generally show much higher cellularity, more numerous blood vessels, and more frequent spindle-shaped cells than aggressive angiomyxomas[1].

In addition to angiomyxoma, a variety of neoplastic and non-neoplastic tumors are found in the non-epididymal extratesticular location[7]. Lesions that contain fat suggest the diagnosis of lipoma, liposarcoma or AMF-like tumor. Fat is present in 24%–56% of cases of AMF-like tumors, but it is not a prominent feature[4]. Other benign mesenchymal lesions include leiomyoma and fibrous pseudotumor. Malignant tumors are less common than benign tumors, the majority being sarcomas that arise from the spermatic cord[7]. Since extratesticular mesenchymal tumors may be benign or malignant, and may occur in patients aged over 60 years, we could not, in our case, exclude the possibility of a malignant tumor.

AMF-like tumors have been described in two previous small series by Laskin *el al*.[4] and Isawa and Fletcher[3], and in isolated case reports, but there have been few reports on the imaging features of this lesion, and very little information about the sonographic appearance. In the report of an AMF-like tumor in the inguinal region, the tumor was found, on contrast enhanced CT, to have a relatively homogeneous appearance with small hypodense foci. On MR imaging of the same case, T1-weighted images showed foci of high signal intensity that were confirmed to be fat on fat-suppressed T1-weighted images. Intense enhancement following gadolinium injection reflects the high vascularity of these lesions[1].

On sonography, AMF-like tumors have been previously reported as having a heterogeneous echotexture and minimal flow on color Doppler imaging[1,5]. In our case, the tumor was a well-circumscribed lesion that was isoechoic and minimally non-homogeneous compared to the testis, with color Doppler flow equal to the flow seen in the testes. Theses imaging features suggest that the lesion was benign, although malignancy could not be excluded. The features of the lesion in our case and in the few previously reported cases, however, are non-specific for the diagnosis or the neoplastic nature of the mass. The similarity in shape, sonographic appearance and blood flow with the testes led us to include polyorchidism in the differential diagnosis.

Conclusion

Angiomyofibroblastoma-like (AMF-like) tumors are rare benign mesenchymal tumors that develop in the inguinal region, scrotum and perineum in men[1], usually in the 5th–6th decade of life. In the presented case, we described the sonographic features and the unusual location of this tumor in the scrotum.

Conflict of interest

The authors do not report any financial or personal connection with other persons or organizations, which might negatively affect the contents of this publications and/or claim authorship rights to this publication.

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