

# Angiomyofibroblastoma-Like Tumor of the Scrotum: A Case Report and Review of Literature

## Skrotumun Anjiomiyofibroblastom Benzeri Tümörü: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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### ABSTRACT

Angiomyofibroblastoma-like tumor is a rare mesenchymal tumor of men. It commonly occurs during the fifth to eighth decades of life and mainly involves the inguinoscrotal region. It is derived from perivascular stem cells and has capacity of lipoid and myofibroblastic differentiation. Histopathologically this tumor in the male genitalia mimics female angiomyofibroblastoma but there are morphological and immunohistochemical differences between these lesions. We report a case of an angiomyofibroblastoma-like tumor that arose in the scrotal region in a 40-year-old man.

**Key Words:** Angiomyofibroblastoma-like tumor, Inguinal canal, Male

### ÖZ

Anjiomiyofibroblastoma benzeri tümör, erkeklerin nadir görülen mezankimal tümörüdür. Sıklıkla yaşamın beş ile sekizinci dekatları arasında oluşur ve başlıca inguinoskrotal bölgeyi tutar. Perivasküler stem hücrelerinden gelişir, lipoid ve miyofibroblastik farklılaşma kapasitesine sahiptir. Histopatolojik olarak bu tümörler kadın anjiomiyofibroblastomasını taklit eder ancak iki lezyon arasında morfolojik ve immünohistokimyasal olarak farklılıklar mevcuttur. Yazımızda 40 yaşında erkek hastada skrotal bölgeden kaynaklanan anjiomiyofibroblastoma benzeri tümör olgusu sunulmuştur.

**Anahtar Sözcükler:** Anjiomiyofibroblastoma benzeri tümör, İnguinal kanal, Erkek

### INTRODUCTION

Angiomyofibroblastoma-like (AMF-like) tumor commonly occurs during the fifth to eighth decades of life and mainly involves the inguinoscrotal region. It is derived from perivascular stem cells and has capacity of lipoid and myofibroblastic differentiation (1). The first study was presented by Laskin WB et al. including 11 cases. The study reported uncharacterized mesenchymal tumors resembling female AMF which arise in the inguinoscrotal region of adult men (2). This entity in male genitalia is exceedingly rare and has been described as its female analog or under the name of male AMF-like tumor (3, 4). Here, we report a case of AMF-like tumor in a 40-year-old man and describe its differential diagnosis.

### CASE REPORT

A 40-year-old man had noticed a painless scrotal mass 1 year prior to the medical consultation. The mass enlarged during this period. Physical examination revealed an elastic, hard, slightly mobile and well circumscribed mass. Doppler ultrasound of the scrotum revealed normal

bilateral testes and a 6×5×3 cm solid mass separate from the testis and epididymis. On magnetic resonance imaging, T1-weighted images showed hypointense and T2-weighted imaging showed hyperintense images. The scrotal mass was excised through an inguinal incision. The mass was well encapsulated and had no relation with the testis and spermatic cord. The cut surface of the tumor measuring 6.5×4.5×2 cm was entirely solid, pale yellowish-white in color and focally gelatinous. Small spots of fresh hemorrhage and several deep-yellow flecks were distributed throughout the cut surface. Microscopically, the tumor was composed of small cells embedded in a prominent myxoid/edematous stroma with hypocellular and hypercellular areas. It contained spindle cells, admixed with a minority of epithelioid-shaped cells. Nuclei were spindle to oval in shape, with finely dispersed chromatin and small nucleoli. Bi-nucleated or multi-nucleated cells were occasionally seen. Spindle cells were separated by fine collagen fibers and abundant edematous background (Figure 1). The vascular component was prominent, haphazardly distributed throughout the tumor and with irregularly thickened walls

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containing fibrinoid or hyalinized material (Figure 2). Perivascular arrangement of tumor cells with focal targeting or whirling pattern was also noted. Mitotic activity ranged from 1 up to 4 mitoses per 50 high-power fields. Atypical mitoses, nuclear pleomorphism, hemorrhage, or necrosis were not observed. Neoplastic cells extended at the surgical resection margin. With immunostains, the tumor cells stained strongly for vimentin and smooth muscle actin (SMA) and were focally stained by desmin (Figure 3). Additional stains were negative for cytokeratin, myogenin, S-100, estrogen and progesterone receptor proteins. Stains for CD34 were also negative in the tumor cells but highlighted endothelial cells. The pathologic diagnosis was angiomyo fibroblastoma-like tumor.

### DISCUSSION

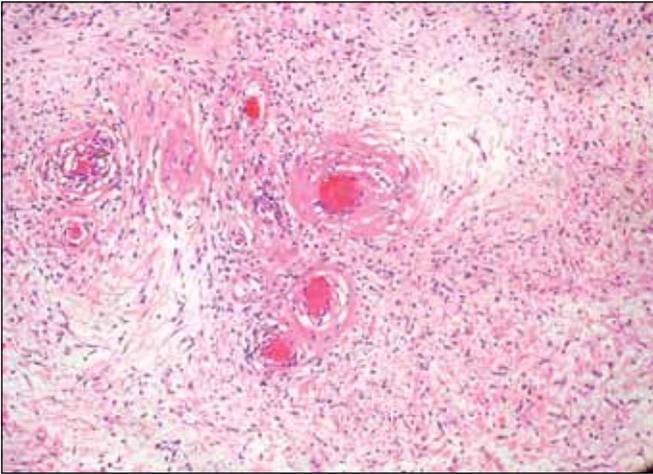
Mesenchymal neoplasms of the genital tract occur predominantly in the vulva, perineum, and pelvis of women but have also been described in men associated with the spermatic cord, inguinal hernias, scrotum, and perineum. Some of these neoplasms occur with or without myofibroblastic differentiation or as a spectrum of spindle cell tumors (5, 6). Although subtle morphologic and immunohistochemical differences between these lesions do occur, they are histogenetically related lesions arising from a common precursor cell (3). One of the mesenchymal neoplasms is AMF that was described by Fletcher CD et al. in 1992 (7). It is a distinct, benign neoplasm that exclusively affects the vulvar region of young to middle-aged female patients. The same tumor in the male genitalia, mimicking female AMF, was reported in 1997 by Nucci MR et al. under the name of cellular angiofibroma (8). This was modified by Laskin WB et al. in 1998 as AMF-like tumor (2). The male neoplasms are superficial, well-marginated masses that are only occasionally associated with pain. Female AMFs are characteristically composed primarily of spindled rather than epithelioid-appearing mesenchymal cells, which proliferate haphazardly or in loose fascicles between vessels without exhibiting a tendency for perivascular growth or forming thin, cordlike configurations or nested aggregates. A condensed collagenous stroma containing a higher concentration of acid mucin prevailed in the majority of male tumors in contrast to the edematous, loosely textured collagenous stroma of the female AMF. Furthermore, elongated, thick-walled vessels and irregularly contoured hemangiopericytomatous vessels commonly accompanied rounded vessels in male tumors. Ectatic or hemangiopericytomatous vessels were only a focal finding in studies of female AMF (1,3,4,9). Perivascular fibrosis is a common feature, and small amounts of intralesional fat are also a frequent finding in both neoplasms (1,5,10).

Intralesional fat was present in 24% of both female and male cases, and it seemed not to be as prominent or as frequent as suggested in prior smaller studies (3). Additionally, both tumors exhibit an immunoprofile including strong and diffuse vimentin immunoreactivity and variable expression of desmin reactivity. SMA, CD34 and estrogen and progesterone receptor proteins are expressed but there is no expression of S-100 protein (7,11). The immunoprofile of the male neoplasm, although still consistent with myofibroblastic differentiation, varied slightly from the female AMF. Desmin expression was noted in 37.5% whereas the figure is 94% for the female AMF reported in the literature. 50% reacted with SMA, whereas this figure is only 15% for all female AMF. The mean estrogen and progesterone receptor scores were substantially higher in the female AMF (7). The immunoreactivity of neoplastic cells for desmin is a common finding in angiomyo fibroblastoma; it is occasionally desmin negative, especially in examples arising in male patients (1,6). Histogenetically, Laskin WB et al. believes that the male AMF-like tumor, like the female AMF, is derived from a perivascular stem cell with a capacity for fatty and myofibroblastic differentiation governed by hormonal, microenvironmental, and growth factor/cytokine-related influences (2). This progenitor cell may be related to the CD34-positive fibroblast like cell that normally resides around vessels. A CD34-positive fibroblast like cell composes the spindle cell element of spindle cell lipoma. In the male AMF-like tumor, this cell may theoretically lose its ability for CD34 expression with myofibroblastic differentiation (4,7).

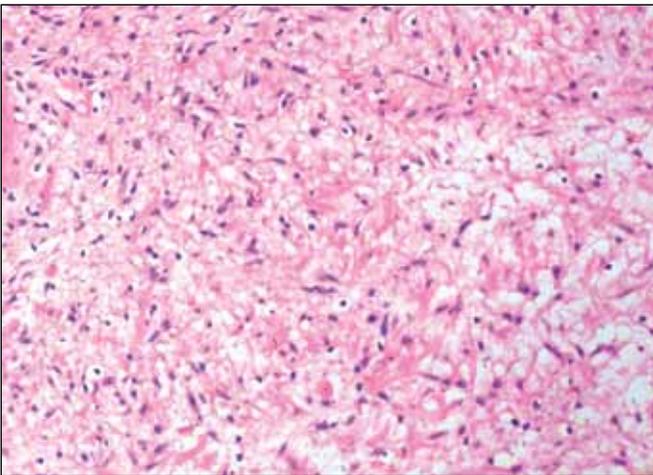
Aggressive angiomyxoma, solitary fibrous tumor, spindle cell lipoma, angiomyo fibroblastoma, schwannoma, hemangiopericytoma, well differentiated liposarcoma and malignant fibrous histiocytoma, are considered in the histological differential diagnosis of AMF-like tumor (1,5).

Aggressive angiomyxoma of the perineal and pelvic soft tissues of women has also been described in the scrotum, perineum, and inguinal region of men. Microscopically, it has an infiltrative growth pattern and a less conspicuous vascular component, which exhibits greater variability in size and distribution of vessels. Small clusters of smooth muscle cells surrounding blood vessels are a characteristic feature of aggressive angiomyxoma. The stromal matrix of the aggressive angiomyxoma is more loosely textured and contains less acid mucin than the stroma of male AMF-like tumors. Tumor cells of aggressive angiomyxoma are immunoreactive for desmin (1,3,5,7).

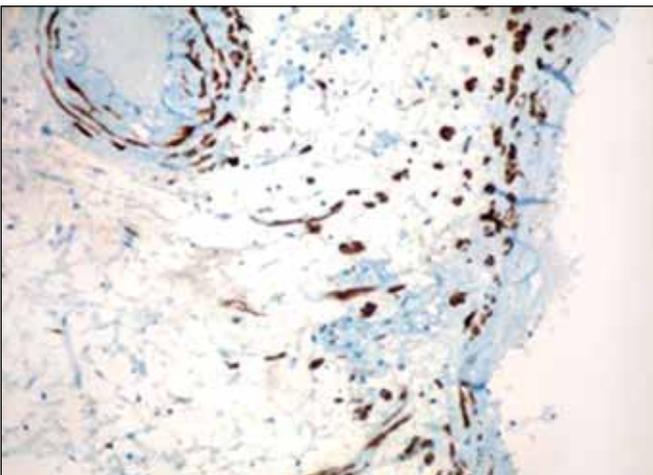
Solitary fibrous tumor has been reported in a wide variety of locations, including the inguinal region, perineum, and



**Figure 1:** Spindle shaped tumor cells proliferating around small to medium-sized vessels within a finely collagenous stroma. Numerous irregular vessels have fibrinoid and hyalinized material in their walls. (H&E, x200).



**Figure 2:** A less cellular area highlights the fibrillary stromal collagen. (H&E, x200).



**Figure 3:** Perivascular spindle cells exhibit cytoplasmic reactivity for desmin (x200).

tunica vaginalis of the testis. Microscopically, solitary fibrous tumor is typically well circumscribed and shows a patternless spindle cell proliferation of alternating hypocellular and hypercellular areas, often associated with dense hyaline collagen bundles, stromal hyalinization, and hemangiopericytoma-like vessels. Angiomyofibrosarcoma-like tumors lack the keloid-type collagen and hemangiopericytoma-like areas that are distinctive of solitary fibrous tumors. The tumor cells of solitary fibrous tumor may also focally express desmin and actin, but they usually exhibit a stronger and more diffuse staining for CD34 (1,3,6,7).

Spindle cell lipoma overlaps AMF-like tumor in several clinicopathological features, including subcutaneous location, age distribution, circumscription, and the presence of spindle cells and adipose tissue. However, most spindle cell lipomas arise in the posterior neck, shoulder, and back regions. Spindle cell lipoma consists of mature adipocytes, bland spindle cells, and short bundles of brightly eosinophilic rope collagen. The blood vessels are usually inconspicuous, whereas the lesion is composed of wispy collagen fibers and numerous small-to medium-sized thick-walled vessels are present (1,3, 10).

Neurilemmoma (benign schwannoma) and neurofibroma typically possess tumor cells with more irregular nuclear contours than the neoplastic cells composing the male AMF-like tumor. Neurilemmoma characteristically demonstrates an alternating pattern of cellular Antoni A foci with hypocellular Antoni B areas. The growth pattern of cells in neurofibroma may be haphazard like that of the male AMF-like tumor, but the former lesion is less vascular (7). Although both peripheral nerve sheath tumors may express CD34, they more commonly express S-100 protein (12).

Hemangiopericytoma features greater vessel density and more variability in vessel size and shape than the vessels in male AMF-like tumors. The lesional cells in hemangiopericytoma are also typically less spindled. Although the neoplastic cells of both lesions may express CD34, the tumor cells of hemangiopericytoma typically lack desmin expression (7).

Liposarcomas, particularly the spindle cell variant of well-differentiated liposarcoma and myxoid liposarcoma, are the most important differential diagnoses clinically as liposarcomas require wide resection. Liposarcoma commonly arises in deep soft tissue, whereas AMF-like tumor arises in subcutaneous tissue. The spindle cell variant of well-differentiated liposarcoma arises superficially and consists of spindled cells and adipose tissue. It differs from AMF-like tumor because of the presence of lipoblasts (1).

Low-grade myxoid malignant fibrous histiocytoma (myxofibrosarcoma) differs from the tumors in our study by exhibiting a more infiltrative growth pattern and cytologically atypical spindled and stellate cells, often accompanied by an intricate network of curvilinear vessels (5,13).

The recommended treatment is wide excision with tumor-free margins, close postoperative monitoring, and long-term follow-up exams. Any suspected recurrence should be imaged and explored. Radiation and chemotherapy play no role in the treatment of this tumor (3).

In conclusion, angiomyofibroblastoma and AMF-like tumors are rare, distinctive, slow-growing, potentially recurrent mesenchymal neoplasms of the female vulva, perineum, or pelvis and male scrotum or inguinal canal. The diagnosis of AMF-like tumor is based on several findings such as superficial location, well-marginated nature of the tumor, spindle bland epithelial cells, numerous small and medium sized vessels with fibrinoid and hyalinized walls set in an edematous collagenous stroma, and low mitotic activity.

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