



Case Report

Angiomyofibroblastoma-Like Tumour of the Scrotum, a Case Report in a Child

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Abstract

Extratesticular solid and cystic masses are commonly encountered in pediatrics. They can lead to a degree of anxiety when discovered by the parents or the caregivers. One of the differential diagnosis is a paratesticular tumour. These are rare mesenchymal lesions. We report this case of Angiomyofibroblastoma (AMF)-like tumour in a child, along with literature review.

Keywords: Angiomyofibroblastoma -like tumour; Cellular angiofibroma

Case Presentation

A 16-month-old healthy boy was brought by his parents to the urology clinic with 1-month history of two painless scrotal swellings. Clinical examination showed 2 small subcutaneous cystic lesions in the left side of the scrotum. The lesions were completely separate from the testis. He was initially treated as possible inflammatory or allergic reaction with non-steroidal anti-inflammatory drugs (NSAIDs) and topical antibiotics.

After 1-month, the child was brought back for a review. The lesions did not respond to the initial treatment. Their presence was confirmed with an ultrasound scan. It reported two subcutaneous hypoechoic heterogeneous mass lesions in the left side of the scrotum, demonstrating prominent flow. They remained stable after a one-month follow-up scan, measuring 1.7 x 0.9 x 0.6 cm and separate from normal testes (Figure 1). Surgical excision with an ellipse of overlying skin was carried out under general anesthesia (Figure 2).

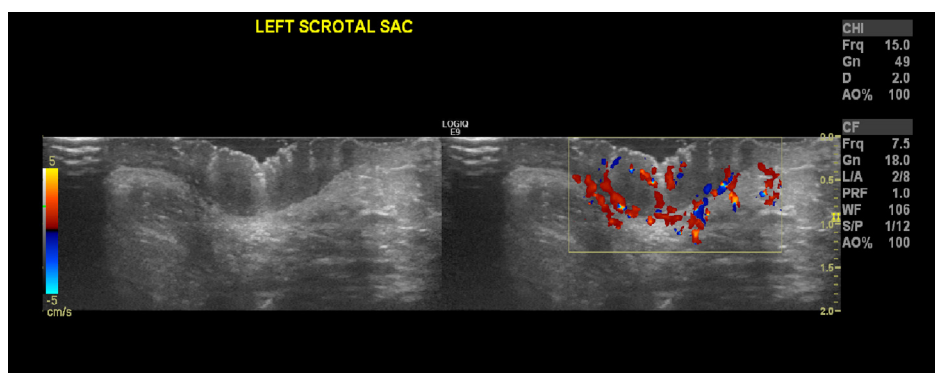


Figure 1: Ultrasound scan showing 2 heterogenous, hypoechoic lesions in the left side of the scrotum and demonstrating prominent flow in the colour doppler image.



Figure 2: Gross appearance of the freshly excised lesions.

Histopathological examination revealed that the two lesions shared the same cellular pattern. It comprised well-circumscribed un-encapsulated lesions characterized by spindle cells lying in an eosinophilic stroma. Occasional stellate and multi-nuclear forms were also present. No mitosis or necrosis is seen (Figure 3). The lesion extends close to the inked margins, but removal appears just about complete.

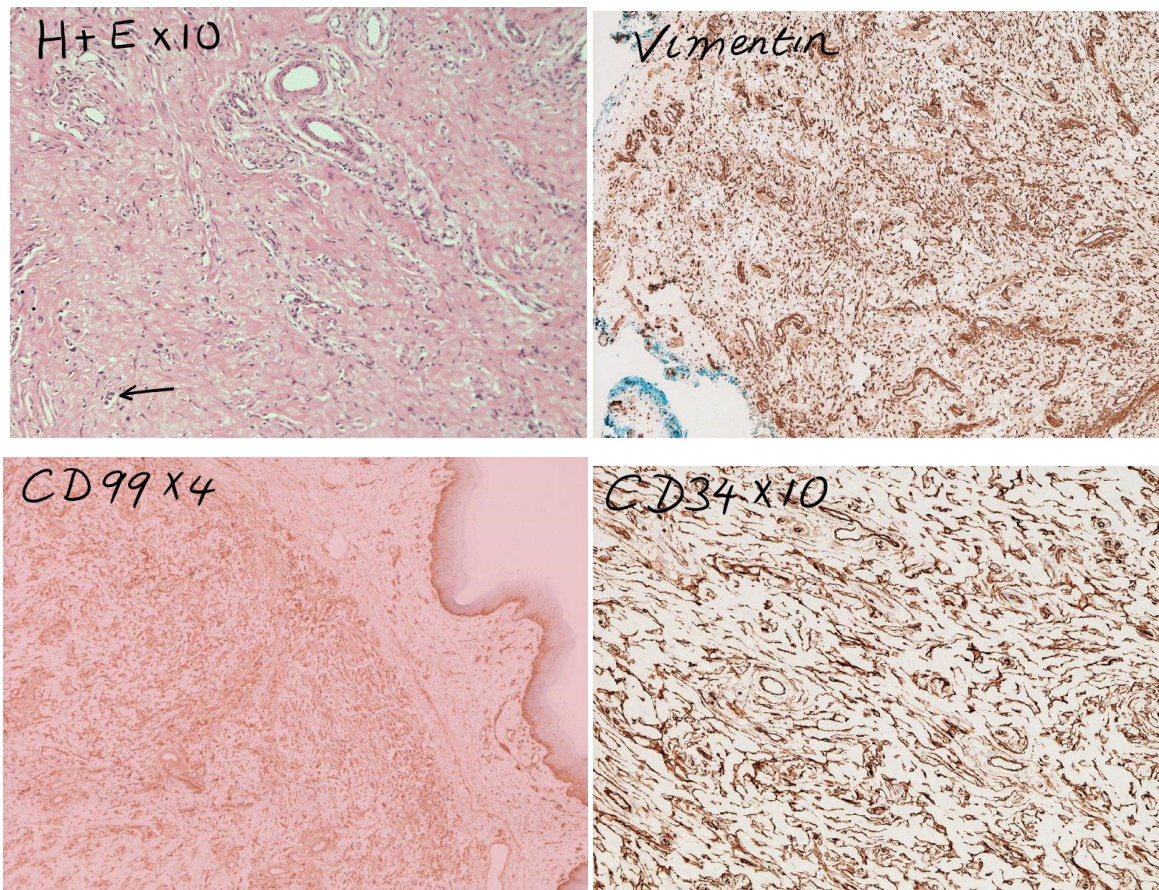


Figure 3: Histopathology and immunohistochemistry slides: **a.** H+E stain showing spindle cells lying in an eosinophilic stroma. Occasional stellate and multi-nuclear forms (arrow) were also present. **b.** Positive Virmetin stain. **c.** Positive CD99 stain. **d.** Positive CD34 stain.

Immunohistochemistry showed positive staining of the spindled cells with vimentin, CD34 and CD99 and focal weak staining for bcl-2. There was negative staining for S100, SMA, cytokeratin, CD31 and desmin. The patient was followed up for 18 months.

Clinical examination showed a healed scar without evidence of tumor recurrence.

Discussion

Angiomyofibroblastoma (AMF) is a rare benign mesenchymal tumor that occurs predominantly in the female external genitalia (vulva, vagina, pelvis and perineum). AMF was first described by Fletcher et al in 1992 [1]. It is a rare tumour that occurs in the reproductive system of middle aged women [1]. AMF-like tumour was first described as a cellular angiofibroma [2]. Thereafter, in 1998, Laskin et al. termed it an AMF-like tumour [3]. In males, AMF-like tumours are known to occur in areas such as the inguinal area, scrotum and perineum [4]. The majority of the cases of AMF-like tumour reported in the literature are in adults. This paper reports one case in a young child and offers discussion of some aspects of its clinical and pathological features. The male AMF-like tumour is a soft tissue neoplasm of the male genital tract that shares the clinicopathological features and proposed perivascular stem cell derivation with both the female AMF and spindle cell lipoma [3].

In the largest series to date, Iwasa and Fletcher reported 25 cases involving men. The age range was 43 to 78 years with a mean age of 52 years. The lesions ranged in size from 0.6 cm to 25 cm with a mean size of 6.7 cm [5]. The most common sites are the inguinal region and the scrotum. AMF-like tumour in males typically follow benign clinical course with only a few reports of invasive or recurrent disease [3,6]. The youngest adult case of AMF-like tumour was reported by Bouhajja et al in 2017 in a 27-year-old patient [7]. Chami et al. reported a case of benign mesenchymal tumour with myofibroblastic differentiation in a 9-year-old girl arising in the left groin that met the histologic features described for myofibroblastoma in adults [8]. Treatment is surgical resection with wide excision to tumour-free margins to prevent recurrent disease. At gross pathological analysis, AMF-like tumours are superficial and well marginated. The tumour cut surface is soft to rubbery and the colour is grey white to yellow brown [9]. Histologically, AMF-like tumour have characteristic prominent vessels with perivascular hyalinization. Tumour cells are spindle cells with limited atypia and low mitotic activity [10]. Small quantities of fat is present in 24% to 56% of cases [3,5]. Immunohistochemical staining is positive for vimentin, variably positive for desmin, muscle specific actin and CD34 and negative for S100 [11]. The differential diagnosis includes a number of uncommon lesions including liposarcoma, spindle cell lipoma, solitary fibrous tumour, leiomyoma and aggressive angiofibroma [12]. The presence of well-defined margins may suggest AMF-like tumour over aggressive angiofibroma which typically displays an infiltrative pattern of growth.

Conclusion

Rare mesenchymal lesions of the scrotum such as AMF-like tumour can be expected in all age groups. Wide surgical excision is the treatment of choice followed with long term follow up, although recurrent disease is very unlikely.

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