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Incidental resection of a scrotal aggressive angiomyxoma mimicking a spermatocele: a case report

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Summary
Background Aggressive angiomyxoma is a rare mesenchymal neoplasm, occurring mainly in females. It is located in the pelvis and perineum, with known metastasis and hormone sensitivity only in females. Local recurrence is relatively common. We describe the case of a 62-year-old man who presented with symptoms and signs of a spermatocele.
Methods Scrotal exploration with surgical excision of the lesion was done. Because a benign setting was assumed, no radical inguinal orchiectomy was performed.
Results The specimen sent to pathology confirmed an aggressive angiomyxoma with positive resection margins.
Conclusion Despite the plurality of benign scrotal masses such as spermatoceles or hydroceles, rare neoplasms should always be kept in mind. Hence, complete excision should be performed whenever possible. We selected an active surveillance strategy despite positive margins, since there is no described case of metastasis in men to date. Therefore, regular scrotal ultrasound examinations every 3 months were arranged as follow-up.

Keywords Aggressive angiomyxoma · Scrotal · Male · Surveillance strategy · Surgical resection

Main novel aspects

- Due to the fact that an ordinary benign setting was assumed, a scrotal exploration adequate for a spermatocele was performed, resulting in positive resection margins.
- Nevertheless, we decided on an active surveillance strategy with fine-meshed ultrasound follow-up, performed by a dedicated uroradiologist.
- However, no signs of local recurrence were detected in the following 6 months.

Background

Aggressive angiomyxoma (AAM) is a rare mesenchymal neoplasm, occurring in the pelvis and perineum mostly in females [1, 2]. It was first described by Steeper and Rosai as a distinctive type of gynecological soft tissue neoplasm [3]. Generally, cystic lesions in the scrotum are common and mostly benign. They may be located in the testis, epididymis, and spermatic cord or they may be a hydrocele. Possibilities in the epididymis include tubular ectasia, cysts in the head or appendix testis, appendix epididymis, or a spermatocele. Aggressive angiomyxomas in males are rare. Only a few cases in males have been described, with less than 20 of them mentioning a scrotal genesis [1, 2, 4]. Aggressive angiomyxoma is described as a locally aggressive neoplasm [5] with a high tendency to recur [6]. We describe the case of a man presenting clinically with a spermatocele, which even

Availability of data and material The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

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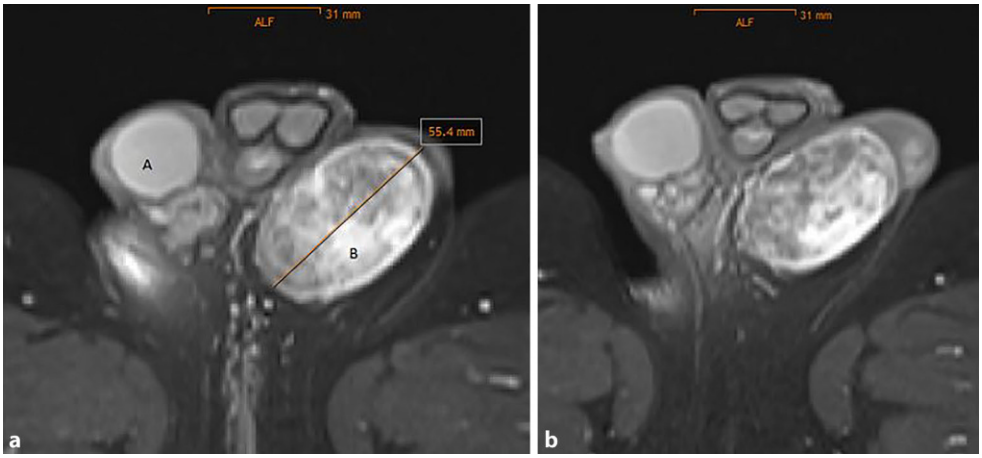

Fig. 1 Magnetic resonance tomography images of the scrotal mass. **a** T1-weighted image of the right testicle (*A*) and the scrotal mass with a length of 55 mm (*B*); **b** T2-weighted image of a different tomographic section

at surgical exploration was not obvious to be a neoplasm. Dealing with this kind of neoplasm is empirical, as currently no guidelines exist.

Case presentation

A healthy-looking non-smoking 62-year-old man presented with a painless, mobile, and soft mass cranial to the left testis. The clinical diagnosis of a spermatocele was made. On ultrasound, a cystic lesion was identified without any signs suggestive of malignancy. Magnetic resonance tomography showed a cystic lesion with some contrast enhancement to its lateral aspect, a finding that conforms to a spermatocele. No lymphadenopathy was shown. The size of the lesion disturbed the patient and therefore a surgical resection was desired (Fig. 1).

An exploration and macroscopically total surgical excision using a scrotal skin incision followed. Contrary to a spermatocele, the 4 × 5 × 7-cm big lesion had no connection to the testis or epididymis. The mass was removed in toto. Finally, the histological examination confirmed an AAM with a positive resection margin. This rare diagnosis was made as a result of the typical localization in the pelvic region combined with characteristic histological features such as fibrosclerotic vessel walls and bundles and fascicles of eosinophilic smooth muscle cells, the so-called Catherine wheel. Immunohistochemistry frequently shows actin and desmin reactivity (Fig. 2).

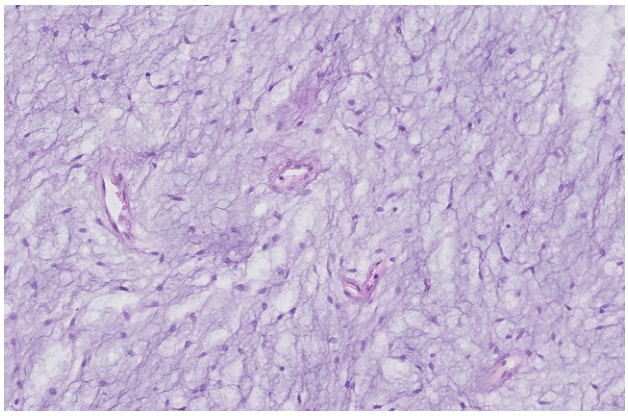

Fig. 2 Histological slice. Aggressive angiomyxoma. Tumor with low cellularity, composed of spindled and stellate-shaped cells within a myxoid-appearing, edematous stroma with a loose meshwork of collagen fibers and thin-walled vessels. H&E, ×200

Discussion

Aggressive angiomyxomas are mesenchymal neoplasms mainly in females arising from the soft tissues in the pelvis and perineum. They are locally infiltrative with recurrence rates varying from 36 to 72% [1, 7]. Two cases with metastasis in females have been described to date [1]. Sensitivity to neoadjuvant and adjuvant hormonal manipulation of estrogen and progesterone has been suggested [1, 8]. Contrary to expectations, a long-term follow-up study did not differentiate between outcomes after complete or incomplete primary resection [9]. As reliable follow-up is possible with our patient, we decided, after informed consent including radical orchiectomy, to perform check-ups every 3 months for the first year. Imaging is done by scrotal ultrasonography by a dedicated uroradiologist. Even a faint suspicion would lead to surgical exploration and radical orchiectomy to facilitate complete resection. If no recurrence is demonstrable, follow-up visits could be stretched out. The patient was free of recurrence with a follow-up of 6 months.

Conclusion

In summary, the etiology and pathophysiology of AAM are poorly understood. The literature suggests slightly different presentations with metastasis only described in females. It is important to realize that cystic lesions in the scrotum do not include only hydroceles or spermatoceles. A high index of suspicion should be maintained during scrotal exploration, demanding

histological analysis to confirm rare findings. Furthermore, complete excisions should be done whenever possible, to attain tumor-free resection margins during primary resection.

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Compliance with ethical guidelines

Conflict of interest M. Kafka, P. Rehder, H. Maier, and W. Horninger declare that they have no competing interests.

Ethical standards A written consent for publication was received.

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