Case: Pediatric paratesticular soft tissue perineurinoma — A rare entity

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Introduction

Soft tissue perineuriomas are rare but benign tumours that are composed of peripheral nerve sheath and are most commonly located in the extremity. Diagnosis is confirmed by the immunohistochemical and ultrastructural features of the tumour. Paratesticular location is extremely rare; so far, only one case is reported. Herein, we report a second case of perineurioma in a 16-year-old male in the paratesticular region.

Case report

A previously healthy 16-year-old boy was referred to urology clinic for a right scrotal swelling. The patient denied any history of trauma, sudden-onset pain, or history of undescended testis. The mass was present and gradually increased in size over three years until it was detected by a nurse at boot camp.

Palpation of the right testicle confirmed that there was a smooth and non-tender mass. An ultrasound revealed diffuse microliths throughout. The right testicle measured 10.7 cm x 6.6 cm x 6.7 cm with a calculated volume of 245.6 mL, normal blood flow to the testicle and associated moderate-to-large hydrocele. (Fig. 1). Serum tumour markers were obtained: normal lactate dehydrogenase (LDH) 380 units/L, beta human chorionic gonadotropin (βhCG) <0.05 units/mL, and alpha-fetoprotein (AFP) was 1.3 ng/mL, all within normal limits. A right radical inguinal orchidectomy was performed.

Gross pathology of the right testis (500 g) and attached spermatic cord revealed a rough, gray-white tunica vaginalis and an unremarkable, gray-white tunica albuginea. Cut sections of the testis revealed white-tan, edematous, and whorled tissue with pinpoint focal areas of hemorrhage. Microscopic evaluation of the mass revealed a well-circumscribed paratesticular mass (Fig. 2). Slender spindle cells arranged in arcs in a myxo-fibrillary background with dystrophic calcifications distributed throughout were identified (Fig. 3). Immunohistochemistry was positive for epithelial membrane antigen (EMA) (Fig. 3A), vimentin, CD34 (Fig. 3B), smooth muscle actin (Fig. 3C), and claudin 1. It is important to note that the tumour stained negative for myogenin, myoD1, desmin, S-100, nuclear beta-catenin, and MUC4. The pathological final diagnosis was myxoid perineurioma.

Discussion

The myxoid presentation of this tumour led to the consideration of a low-grade fibromyxoid sarcoma (LGFMS), but the immunophenotypic profile ultimately supported the diagnosis of a perineurioma. Perineuriomas are rare, benign tumours composed of peripheral nerve sheath that are characterized by their immunohistochemical and ultrastructural features. EMA expression is an important marker that distinguishes perineuriomas from any morphological mimickers and were present in all 81 cases of a clinopathological analysis.[1,2] CD34 and claudin-1 further confirm the diagnosis, while the absence of desmin and S-100 are equally as significant.[1,3] EMA stains are positive in both perineuriomas and LGFMS, but a negative MUC4 stains excludes LGFMS because it is a very specific and sensitive marker for LGFMS.[4,5] Grossly, perineuriomas have a characteristic focally whorled growth pattern, and histologically, elongated and thin spindle cells can be seen.[1]

While most perineuriomas are soft tissue perineuriomas, the location of this perineurioma is what made this case unique. This case is the only other one we know, besides one mentioned in the "Clinicopathologic analysis of 81 cases," of a paratesticular myxoid perineurioma, but no details about that one case were given.[2] The best course of action in such a case, to our knowledge, is to carefully excise the mass so that it doesn't impede any normal testicular function. If more cases are found, they should be compared to look for any similarities or differences so that we can better understand how this type of tumour comes to be in the paratesticular region.

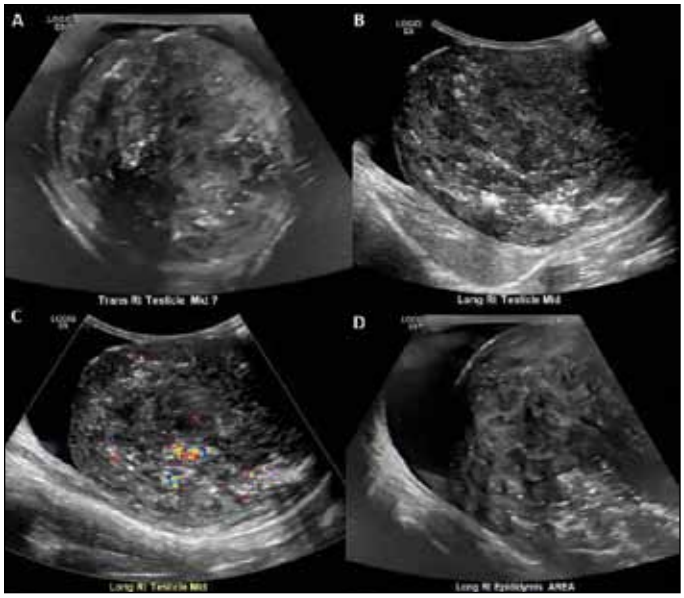

Fig. 1. (A–D) Diffusely enlarged right testicle with heterogeneous appearance and diffuse microliths, measures 10.7 cm x 6.6 cm x 6.7 cm in size.

Conclusion

Perineuriomas are rare, benign tumours composed of perineurial cells, but the occurrence of one in the paratesticular region makes this case even more uncommon. Herein, we report the second incidence, to our knowledge, of a paratesticular myxoid perineurioma and believe surgical excision is the best treatment plan in this situation.

Competing interests: The authors report no competing personal or financial interests.

This paper has been peer-reviewed.

References

1. Macarenco RS, Ellinger F, Oliveira AM. Perineurioma: A distinctive and under-recognized peripheral nerve sheath neoplasm. *Arch Pathol Lab Med* 2007; 131:625-36.
2. Hornick JL, Fletcher CD. Soft tissue perineurioma: Clinicopathologic analysis of 81 cases, including those with atypical histologic features. *Am J Surg Pathol* 2005;29:845-58. https://doi.org/10.1097/01.pas.0000155166.86409.d2
3. Chetty R. Myxoid perineurioma presenting as a gastric polyp. *Ann Diagn Pathol* 2010; 14:125-8. https://doi.org/10.1016/j.anndiagpath.2009.06.002
4. Yang EJ, Hornick JL, Qian X. Fine-needle aspiration of soft tissue perineurioma: A comparative analysis of cytomorphology and immunohistochemistry with benign and malignant mimics. *Cancer Cytopathol* 2016;124:6518. https://doi.org/10.1002/cncy.21725
5. Ban L-K, Tseng AH, Huang S-H, et al. Low-grade fibromyxoid sarcoma of the external anal sphincter: A case report. *World J Surg Oncol* 2017;15:109. https://doi.org/10.1186/s12957-017-1174-8

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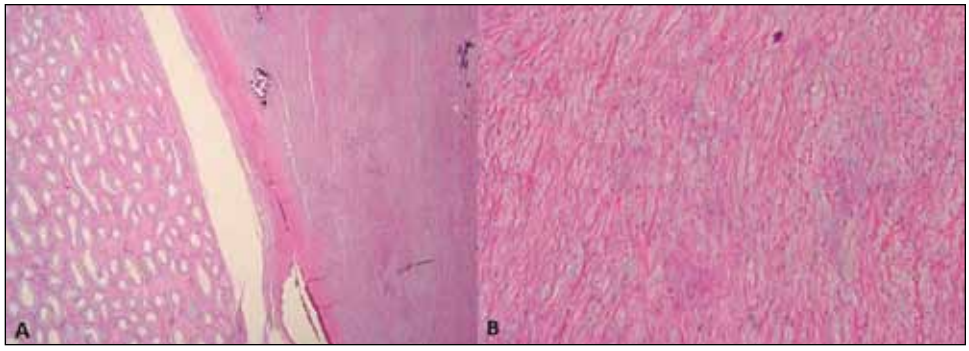

Fig. 2. (A) Well-circumscribed paratesticular mass on the right in relation with the testicular tissue on the left. ***(B)*** The tumour consists of slender spindle cells arranged in arcs in a myxo-fibrillary background with dystrophic calcification.

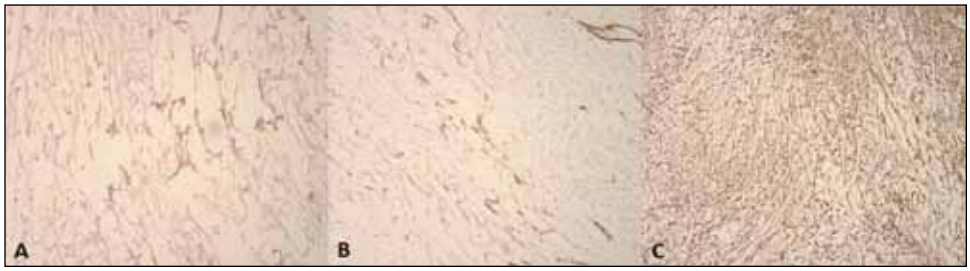

Fig. 3. (A) Tumour cells are positive for epithelial membrane antigen. ***(B)*** Tumour cells are positive for CD34. ***(C)*** Tumour cells are positive for smooth muscle actin.