

Intrascrotal extratesticular schwannoma: A first pediatric case

Michelle Bergeron,* Stéphane Bolduc, MD, FRCSC; Sébastien Labonté, MD, FRCPC;†
Katherine Moore, MD, FRCSC*

*Department of Surgery, Division of Urology, CHU de Québec, Université Laval, Québec City, QC; †Department of Molecular Biology, Clinical Biochemistry and Pathology, CHU de Québec, Université Laval, Québec City, QC

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Abstract

Scrotal nerve sheath tumours unassociated with neurofibromatosis or schwannomatosis are extremely rare. Very few cases of benign intrascrotal and extratesticular schwannomas have been reported, but none of them occurred in childhood. This current report describes for the first time a case of benign intrascrotal extratesticular solitary schwannoma in a 16-year-old male.

Paratesticular tumours are infrequent neoplasms of mesenchymal origin that can affect the spermatic cord region, testicular tunics, epididymis and vestigial remnants. The most common tumours that arise from the paratesticular tissue are benign neoplasms, such as leiomyoma, lipoma and adenomatoid tumour.¹ Although schwannoma is the most common tumour of the peripheral nerves, it is extremely rare in the paratesticular region. To our knowledge, this case is the first of a benign intrascrotal extratesticular schwannoma in the pediatric population.

Case report

A 16-year-old male presented with a painless and slowly growing scrotal mass first noticed in the past few years. The patient was initially referred to the pediatric urology department with a suspected left varicocele. Otherwise, he had no noteworthy medical or family history, and there was no trauma or infection reported. On physical examination, scrotal palpation revealed an irregular, firm, non-reducible and non-transilluminating mass of about 8 cm in diameter with a mid-scrotal location at the base of the penis (Fig. 1). The mass was distinct from the testes and scrotal wall and did not seem to be attached to any underlying structures. Spermatic cords and scrotal skin were normal. Testes were normal in size and shape. There was no palpable lymph node upon physical examination. Furthermore, there were

no clinical stigmas or family history related to neurofibromatosis or schwannomatosis.

Ultrasonographic examination revealed a solid voluminous, heterogeneous and lobulated mass measuring 4.3 × 4.1 × 5.5 cm in size on the inferior-anterior side of the base of the penis (Fig. 2). The mass was highly vascularized on Doppler examination. Testes and epididymis were normal, except for a cyst or spermatocele of 8 mm in diameter located on the right epididymis, with no signs suggesting a hydrocele. Laboratory results showed β -human chorionic gonadotropin, alpha-fetoprotein and lactate dehydrogenase levels were within normal range. A complete surgical excision of the scrotal mass was performed and did not reveal any particular attachment to adjacent structures except for the vessels at its base. Macroscopic examination of the specimen revealed a well-circumscribed rounded encapsulated mass of 5.5 × 5.0 × 3.5 cm in size and 58.2 g in weight. Its cut surface consisted of a grayish-white fasciculated tissue with very few brownish spots and no obvious area of necrosis or recent bleeding. The histopathological report showed an arrangement of spindle cells in a storiform pattern with Verocay bodies interspersed with myxomatous areas. Blood vessels with hyalinised walls were noted. Immunohistochemistry studies showed diffusely positive S100 protein and immunoreactive perineural capsule with epithelial membrane antigen (Fig. 3). Therefore, we settled on the final diagnosis of intrascrotal extratesticular schwannoma. Afterwards, the patient was assessed by a neurologist and a geneticist, who both concluded that the patient did not present any signs related to neurofibromatosis or schwannomatosis.

Discussion

Schwannomas, also named neurilemmomas or neurinomas, are benign tumours of the nerve sheath that develop from myelin-forming cells called the Schwann cells. They mostly affect the head and neck areas along with the nerves of the flexor surfaces of the upper and lower extremities, with a

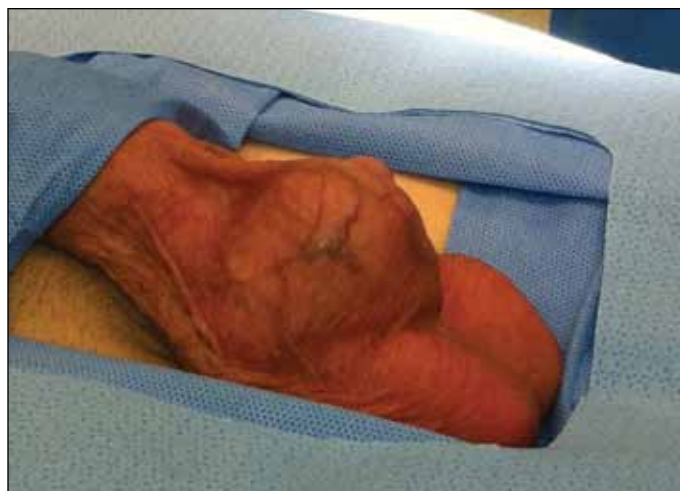


Fig. 1. Mid-scrotal mass seen in a 16-year-old male.

peak patient age between 20 and 50 years old.² Multiple schwannomas can be seen as part of type 2 neurofibromatosis, schwannomatosis or Carney complex. Schwannomas can cause symptoms of dysesthesia, sensory loss, weakness or radicular-type pain, but most patients will remain asymptomatic.³

Histologically, schwannomas are encapsulated tumours composed of cells arranged in a biphasic architecture made of Antoni A and Antoni B areas. Antoni A areas refer to dense bundles of spindle cells with palisading nuclei (Verocay bodies). Antoni B areas are characterized by loose-textured tissue, fewer cells and myxoid changes.⁴ Immunohistochemistry studies of schwannomas show a uniform staining with S100 protein.⁴ Malignant transformations are extremely rare.⁵

Although schwannoma is the most common tumour of the peripheral nerves, it represents a rare finding in the differential diagnosis of scrotal neoplasms and paratesticular lesions. Few cases of genital schwannomas, involving the prostate, spermatic cord, testis, penis, tunica vaginalis and seminal vesicle, have been reported.⁶⁻⁷ Based on our literature review, 10 previous reports of solitary benign intrascrotal extratesticular schwannomas unassociated with neurofibromatosis or schwannomatosis involved patients ages between 24 and 71.⁸⁻¹¹ To our knowledge, this is the first pediatric case of

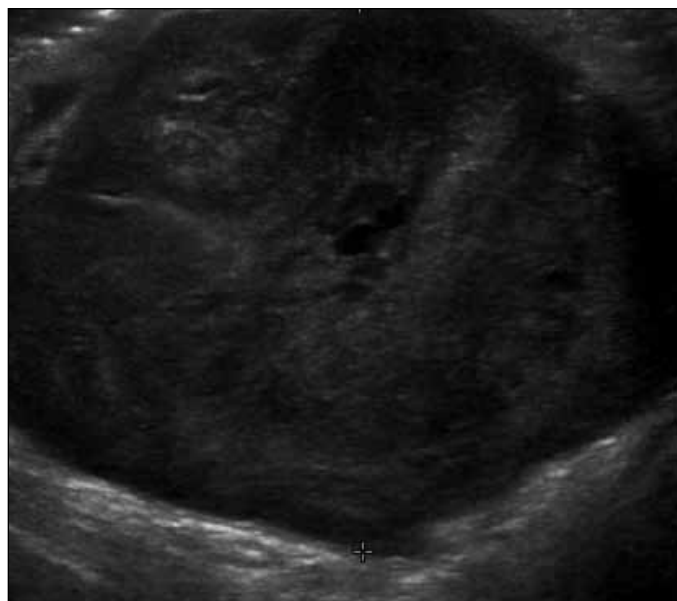


Fig. 2. Ultrasound imaging showing an intrascrotal extratesticular mass located at the base of the penis.

benign intrascrotal extratesticular schwannoma. However, extremely rare childhood cases of neurofibroma, another benign nerve sheath neoplasm, and schwannomas of the clitoris, have been reported.¹²⁻¹³ Schwannomas' slow growth and usually asymptomatic presentation could explain the infrequency of these tumours in the pediatric population.

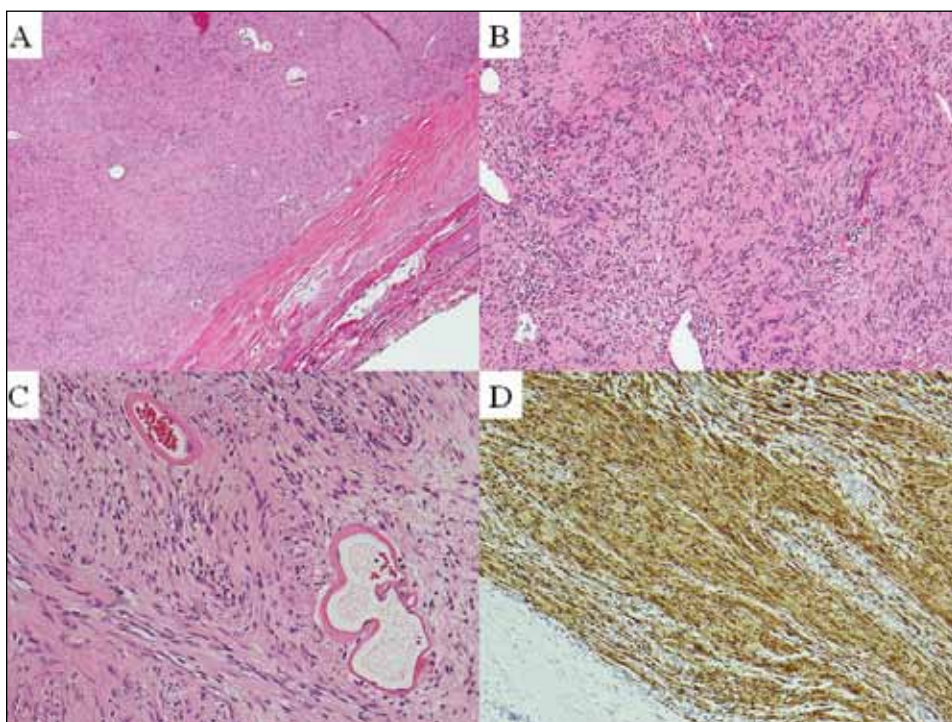


Fig. 3. A: Well-circumscribed tumour with fibrous capsule. B: Proliferation of spindle cells with formation of Verocay bodies. C: Short fascicles of bland spindle cells with hyalinized blood vessels. D: Strong diffuse immunoreactivity for S100.

In the reviewed cases, acute pain or occasional tenderness was uncommon. Ultrasonographic examination was performed in all cases and therefore seemed to be the gold standard in the evaluation of scrotal masses, while computed tomography or magnetic resonance imaging were rarely performed. Complete surgical excision of the tumour was executed in all reviewed cases with favourable results. There was no recurrence noted with any of the benign intrascrotal extratesticular schwannomas. However, periodical surveillance after complete excision was still promoted by some authors.⁹⁻¹⁰

Conclusion

Although it is extremely rare, schwannoma must be considered in the differential diagnosis of a paratesticular tumour in a pediatric patient, along with other more common benign neoplasms of the paratesticular tissue, such as leiomyoma, lipoma and adenomatoid tumours.

Competing interests: Michelle Bergeron declares no competing financial or personal interests. Dr. Bolduc is principal investigator for Astellas. Dr. Labonté has received an honorarium for lectures given and consensus meetings from Merck and Bristol-Myers-Squibb. Dr. Moore is currently an investigator with Astellas in a clinical trial.

This paper has been peer-reviewed.

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Correspondence: Dr. Katherine Moore, Department of Surgery, Division of Urology, CHU de Québec, Université Laval, Québec City, QC; katherine.moore.1@ulaval.ca