

Inflammation and infection

Perineal schwannoma: A case report with novel genitourinary association and histopathology

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ABSTRACT

We describe a 39-year-old male patient who presented with a large perineal mass found to represent an entity rarely seen involving the genitourinary tract, a perineal schwannoma. We also provide a discussion regarding the common presentation, diagnostic approaches and treatment options for patients who present with this rare entity.

Introduction

Schwann cells are peripheral nervous system-myelinating cells of neural crest origin. A schwannoma is typically a benign tumor of peripheral nerve sheath origin although malignant cases have been reported.¹ Although commonly associated with genetic syndromes such as neurofibromatosis type 2, they can also be found in isolation in locations such as the head, neck, and perineum.^{2,3} Perineal schwannomas are quite rare, with largely case report-level descriptions of this pathology.

We describe a 39-year-old male patient who presented with a large perineal mass found to represent an entity rarely seen involving the genitourinary tract, a perineal schwannoma. We also provide a discussion regarding the common presentation, diagnostic approaches and treatment options for patients who present with this rare entity.

Case description

A 39-year-old male presented to clinic with a painless mass that began as a small bump in the perineum and grew steadily over two years. He had no history of fever, weight loss, or urinary tract symptoms. There was no significant past medical, surgical, family, or social history.

On physical exam, there was a firm, non-tender, lemon-sized mass on the midline of the perineum. The testicles were non-tender, palpable bilaterally within the scrotum, without masses or abnormalities, and independent from the perineal mass. Complete blood count, coagulation

profile, and basic metabolic profile were all within normal limits. Serum alpha fetoprotein (4.8 ng/mL), beta human chorionic gonadotropin (<2.0 mIU/mL), and lactate dehydrogenase (189 U/L) levels were obtained as tumor markers and also found to be within normal limits. CT chest, abdomen, and pelvis with contrast demonstrated few mildly prominent nonspecific right inguinal lymph nodes with unremarkable vasculature as well as a heterogeneously enhancing pedunculated mass arising from the perineum, measuring 6.2 × 4.8 × 5.2 cm (Fig. 1). Vascular supply to the mass was seen originating from multiple prominent vessels branching from the right femoral artery. Overlying soft tissues were unremarkable.

In the operating room, flexible cystourethroscopy and excisional biopsy of the perineal mass were performed with no complications. A 6.3 × 5.5 × 5.5 cm specimen with negative resection margins was obtained (Fig. 2). Serial sectioning revealed a well-circumscribed, variegated, focally hemorrhagic mass with 1 cm resection margins. Pathology identified the mass as a benign schwannoma.

The schwannoma was a biphasic nerve sheath tumor; these are composed of a cellular component (Antoni A) often with palisading Verocay bodies and a myxoid component (Antoni B). Our patient's tumor is solitary and grossly well-circumscribed. The cells have ill-defined cytoplasm, are narrow, elongated, and wavy with tapered ends interspersed with collagen fibers. The Verocay bodies show nuclear palisading around fibrillary processes seen in the cellular areas. The hypocellular Antoni B areas contain large, irregularly spaced vessels

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Fig. 1. Axial, Coronal and Sagittal images showing 6 cm heterogeneous perineal mass with vascular stalk.

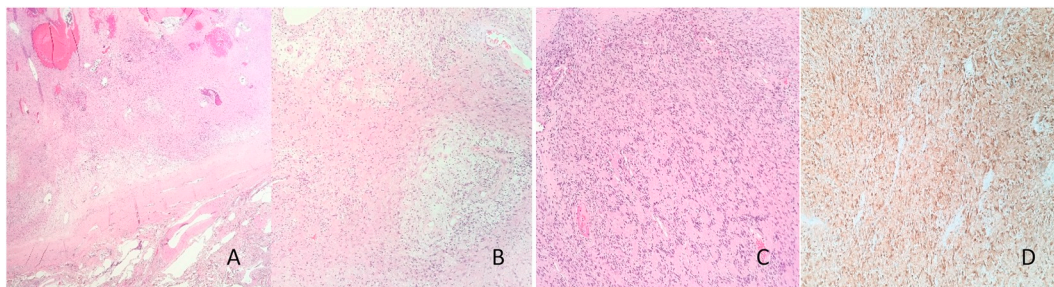


Fig. 2. Figure A shows a well circumscribed tumor, surrounded by fibrovascular connective tissue. The low power view demonstrates Antoni A cellular areas and Antoni B myxoid areas with irregularly spaced vessels. Figure B shows a closer view of both areas. Notice the prominent hyalinization of the vessels in the Antoni B areas. Figure C shows Antoni A cellular areas with prominent palisading, characteristic of Verocay bodies. Figure D shows S-100 protein immunohistochemistry positivity, as expected in a schwannoma.

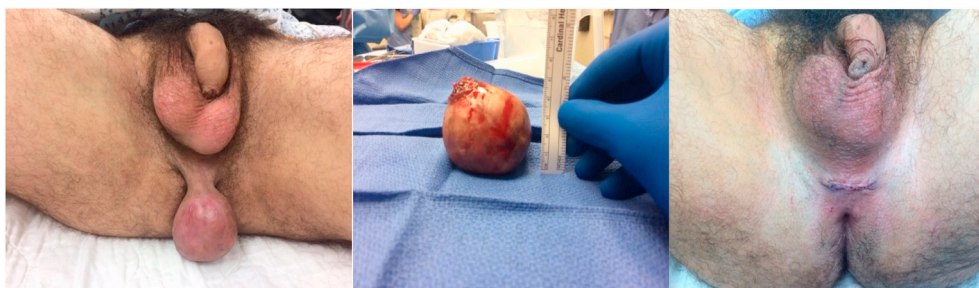


Fig. 3. En-bloc excision of large perineal mass.

with thickened, hyalinized walls (Fig. 3). Pan-keratin, HMB45, and smooth muscle actin immunohistochemical stains were all negative. S-100 and CD68 immunohistochemistry analysis revealed positive staining for these two markers, which supported the diagnosis of schwannoma.

Ten-day and three-month follow-ups in clinic revealed a well-healed incision and normal voiding and sexual function. Physical exam as well as CT chest, abdomen, and pelvis at three months demonstrated no evidence of metastasis or recurrence.

Discussion

Perineal schwannomas are a rare entity typically handled by general surgeons and urologists, depending on the structures involved or surrounding.^{1,3} Typically of benign origin, this pathology often presents as an asymptomatic mass.² Any symptomatology would be based on the degree of involvement in the surrounding structures such as the anus, urethra or nerves in the perineum.

Most cases outline complete excision with negative margins as the treatment of choice due to the tumor's propensity for recurrence, particularly in syndromic and malignant cases.⁴ In our particular case, the pedunculated schwannoma did not pose a threat to surrounding gastrointestinal structures.³ Although our case was not associated with the gastrointestinal tract, care still had to be taken to maintain correct planes during dissection to avoid pelvic floor muscle resection or nerve

damage in the perineum, which could result in subsequent incontinence or erectile dysfunction. While iatrogenic injury could be avoided with the use of a partial resection, the argument against a partial surgery would be a large propensity to recur.³

With the help of pathology, a final diagnosis of schwannoma can be made. In particular, schwannoma has a specific cellular component and a myxoid component as outlined above.⁵ Histopathologically, S-100 positivity supports the diagnosis of schwannoma.⁵ Our particular case yielded an S-100 positive specimen as well as the cell marker CD68 which did not seem to be found in the literature in reference to perineal schwannomas. In our case, a final pathologic diagnosis of benign schwannoma involving the perineum was made.

Conclusion

In summary, perineal schwannomas are a rare pathology often presenting as large masses as seen in our patient. We describe here a case report of massive perineal schwannoma in an adult male with regards to treatment, pathology and follow-up management. Our case is novel in its histopathologic analysis including CD68 as well as its size and close association with genito-urinary structures.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.eucr.2019.101021>.

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