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Perianal Schwannoma: A Rare Case

[1]Dr.Rajni Bhardwaj, Govt. Medical College, Jammu, J & K.

[2]Dr.Deepti Chanjotra, Govt. Medical College, Jammu, J & K.

[3]Dr. Ishiya Anthal, Govt. Medical College, Jammu, J & K.

Corresponding Author: Dr. Deepti Chanjotra, Govt. Medical College, Jammu, J & K.

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Abstract

Schwannoma is a rare type of tumour that forms in the nervous system. Schwannoma grows from cells called Schwann cells that protect and support the nerve cells of the nervous system.These are often benign, but can turn malignant in rare cases. Perianal schwannomas are exceptional and rarely reported in literature. We report a case of a 45 years old female who presented to the Surgery OPD, GMC Jammu, with complaints of recent onset discomfort due to a perianal swelling which started developing a year ago. She was diagnosed as a case of a benign perianal schwannoma which was excised completely to avoid recurrence. Careful dissection was carried out to avoid risk of incontinence. She was followed up in OPD and the postoperative period was uneventful.

Keywords: Schwannoma, Perianal, Tumour, Incontinence, Sphincter, Immunohistochemistry, Histopathology, Biopsy

Clinical Case

A 45 years old female with no known comorbidity presented to the surgery OPD with complaints of a swelling in the perianal region on right side which gradually increased in size over a period of one year. The swelling was painless initially but for last one month, she had discomfort on sitting for longer duration which got relieved on standing and doing her routine activities. There were no complaints of rectal bleeding or bowel disturbance. On examination, she had a firm, oval, smooth swelling with well-defined margins, sized about 7×5 cm in the right perianal region. The swelling was tender to touch, mobile with overlying skin showing early changes of pressure necrosis with minimal excoriation. Digital rectal examination was painless, the rectum showing intraluminal bulge on the right side due to mass effect of the swelling. PV examination was normal. Preoperative tissue biopsy of the swelling showed spindle shaped cells with elongated nuclei without atypia. CT scan pelvis revealed a well-defined

oval iso-dense 92×82×86 mm swelling in the right ischioanal region in the subcutaneous plane without any calcification or haemorrhage in the lesion and the lesion is abutting the right external anal sphincter anterosuperiorly. CEMRI of the pelvis documented oval shaped mass lesion measuring 9.3×9.4×5.1 cm, isointense to muscle causing mass effect with leftward displacement of anal canal and on postcontrast imaging, the lesion shows intense heterogenous enhancement with presence of perilesional collaterals along inferomedial aspect. The patient was planned for excision of the swelling after proper counselling regarding likely injuries to sphincter complex and neurovascular bundle in the proximity of the swelling. The swelling was excised via a parasaggital skin incision made in the perianal region with the patient in prone jack-knife position under spinal anaesthesia. Adequate care was taken to preserve the anal sphincter complex, the nerves and vessels in the proximity of the mass. It was a lobulated, well capsulated mass with multiple collateral vessels and thick fibrous adhesions. The mass was, however, not in contact with any particular muscle or neurovascular structure. Suction drain was kept in the wound cavity and removed on the 4th postoperative day. Postoperative histopathology revealed features of Benign Nerve Sheath Tumour, i.e, Schwannoma. On immunohistochemistry, tumour cells were reactive to S-100 antibody. The postoperative period was uneventful and the patient was discharged on the 5th postoperative day. She was followed up on OPD basis and did not have any complaints regarding the wound or anal sphincter dysfunction.

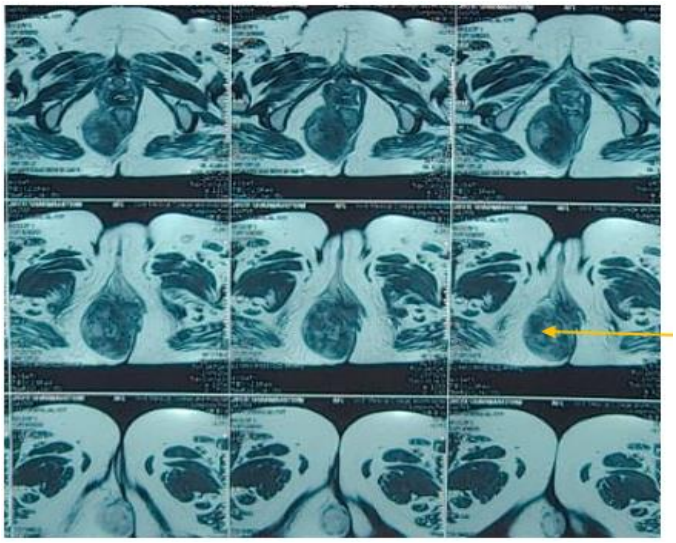

Figure 1: MRI film showing right perianal schwannoma (yellow arrow).

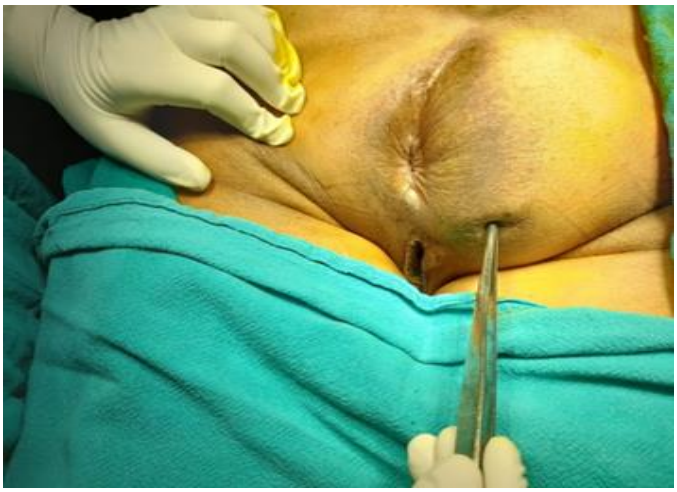

Figure 2: Preoperative location of the swelling.

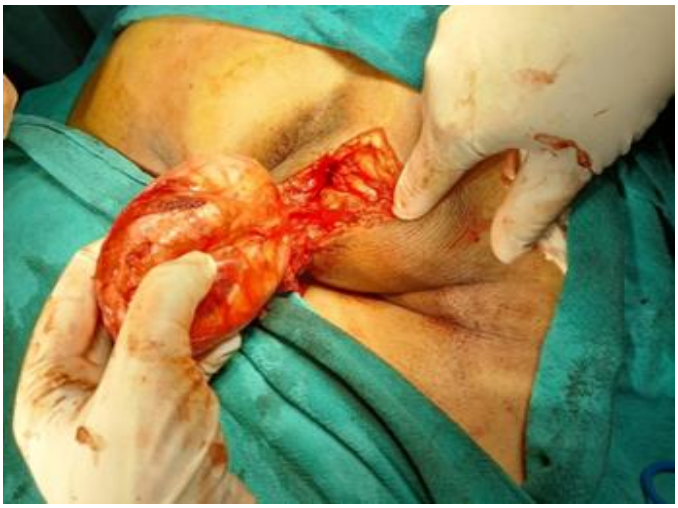

Figure 3: Excision of the mass from its location.

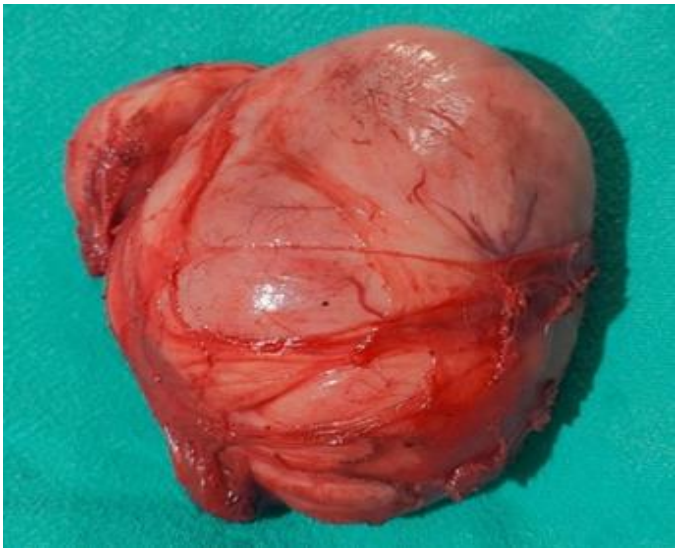

Figure 4: Specimen (lobulated mass) sent for histopathology.

Discussion

Perianal schwannoma is a rare tumour. The rate of reported pelvic schwannomas is only 1%[1]. These are mostly benign and have a very low rate of malignant transformation. Preoperative diagnosis is difficult since both benign and malignant tumours typically present as asymptomatic enlarged perineal mass causing compression on adjacent organs, especially the urethra and rectum.

Benign schwannoma is an encapsulated nerve-sheath neoplasm originating from Schwann cells. These tumours are more frequently located in the head, neck, upper and lower extremities and trunk; retroperitoneal localization is unusual[2] and development in the perianal area is extremely rare. Malignant transformation is rare[3]. These tumours are composed of Schwann cells which support the peripheral nerve fibres and are neuroectodermal in origin. Schwannoma is enveloped by a true capsule consisting of the perineurium of the nerve bundle of origin, surrounded by a condensation of the deepest layers of the epineurium. The capsule is often covered by tortuous blood vessels and allows excision of the tumour without damage to the parent nerve. The rare plexiform variant of a schwannoma may infiltrate between nerve bundles and thus make excision difficult[4]. Schwannomas characteristically form an eccentric oval swelling, with the attenuated nerve bundles of the parent nerve stretched and displaced over the dome of the mass. They grow slowly, but in sites such as the mediastinum and retroperitoneum, may attain a large size before presentation and their true nature may remain unrecognised[5,6]. Such lesions often display extensive degenerative features including the formation of a cyst, fibrosis and calcification. The histomorphological picture of the tumour contains varying proportions of two distinctive tissues, Antoni-A and Antoni-B. There is formation of cysts, hyalinisation of the matrix and focal calcification. An inflammatory infiltrate is usually present including numerous histiocytes, some of which contain haemosiderin. The nuclei of the Schwann cells become hyperchromatic, enlarged and multilobed, but mitoses remain sparse. Although such nuclear atypia may cause concern, the association with other degenerative features and the lack of mitoses indicate a benign tumour. The cellular schwannoma is composed almost entirely of Antoni-A tissue. The spindle cells are closely packed and may be arranged in a herringbone pattern. Nuclear palisading and Verocay bodies are absent. The mitotic rate is low. Staining with S-100 protein is uniformly positive. In contrast, the malignant peripheral nerve-sheath tumours show scattered positive cells or is only focally positive. The infrequency of schwannomas in general clinical practice causes difficulties in diagnosis and delay before recognition[7]. Unnecessary resection of the parent nerve or nerves is a serious complication[4,8].

Accurate diagnosis is essential in the treatment of any tumour. Sondack and Chang[9] state that a properly performed biopsy is the critical first step in a multimodality approach to treatment since if it is done improperly, it can complicate the care of the patient and affect treatment. Diagnosis of a schwannoma arising within a nerve trunk of the upper or lower limb is usually clinical where the swelling is painful to pressure and is mobile from side to side, but not in the vertical axis of the limb. Percussion induces painful paraesthesia in the area of the nerve of origin. A mass arising from a nerve trunk which causes pain and is attended by deepening loss of function is indicative of a malignant tumour. For schwannomas arising from nerve trunks within the limbs, the proper treatment is excision with the object of removing the tumour while preserving nerve function. Biopsy is required when there is any doubt about diagnosis. It is for the responsible surgeon to decide whether biopsy is needed and which method is used. Hence, confirmation of the diagnosis is the first step in definitive treatment.

Conclusion

Pelvic schwannomas are very rare tumours, usually asymptomatic, presenting as large masses. Our patient presented at an early stage of the development of this rare perianal schwannoma showing mass effect without affecting the nerve function. She was diagnosed early, treated in time and did not suffer any functional impairment of the neuromuscular tissue in the vicinity of the schwannoma.

References

1. Panwar P, Kumar S, Singh S, Sriharsha AS, Gupta K. Giant abdominoperineal malignant schwannoma: an unusual presentation and surgical challenge. Case Rep Urol. 2015;11:1–5.
2. Maneschg C, Rogatsch H, Bartsch G, et al. Treatment of giant ancient pelvic schwannoma. Tech Urol 2001;7:296-8.
3. Woodruff JM, Selig AM, Crowley K, Allen PW. Schwannoma (neurilemoma) with malignant transformation: a rare distinctive peripheral nerve tumour. Am J Surg Pathol 1994;18:882-95.
4. Birch R, Bonney G, Wynn Parry CB. The peripheral nervous system and neoplastic disease. In: Birch R, Bonney G, Wynn Parry CB, eds. Surgical disorders of the peripheral nerves. Edinburgh: Churchill Livingstone, 1998:335-72.
5. Abernathey CD, Onofrio BM, Scheithauer B, Pairolero PC, Shives TC. Surgical management of giant sacral schwannomas. J Neurosurg 1986;65:286-95.
6. Hennigan TW, Branfoot AC, Theodorou NA. Ancient neurilemmoma of the pelvis. J R Soc Med 1992;85:416-7.
7. Kehoe NJ, Reid RP, Semple JC. Solitary benign peripheral-nerve tumours: review of 32 years' experience. J Bone Joint Surg [Br] 1995;77-B:497-500.
8. Birch R, Bonney G, Dowell J, Hollingdale J. Iatrogenic injuries of peripheral nerves. J Bone Joint Surg [Br] 1991;73-B:280-2.
9. Sondack VK, Chang AE. Clinical evaluation and treatment of soft tissue tumours. In: Weiss SW, Goldblum JR, eds. Enzinger and Weiss's soft tissue tumours. Fourth ed. St. Louis: Mosby, 2001:21-44.