ORIGINAL RESEARCH PAPER

General Surgery

ANCIENT SCHWANNOMA AS AN EXCEPTIONAL SACRAL SPINAL TUMOUR: A CASE REPORT

KEY WORDS: Ancient Schwannoma, Sacral spine,

Dr. Dinesh Dutt Sharma* Associate Professor And Unit Head, Department Of General Surgery, Dr S N Medical College, Jodhpur, Rajasthan. *Corresponding Author

Dr. Rakesh Bhatnagar Postgraduate Student Department Of General Surgery, Dr S N Medical College, Jodhpur, Rajasthan.

Dr. Hemant Kumar Postgraduate Student Department Of General Surgery, Dr S N Medical College, Jodhpur, Rajasthan.

Dr. Kunal Chitara Postgraduate Student Department Of General Surgery, Dr S N Medical College, Jodhpur, Rajasthan.

ABSTRACT

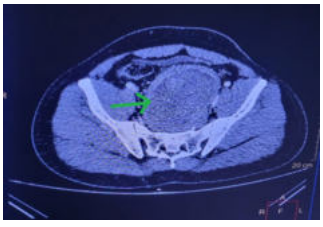
Schwannomas predominantly are non-cancerous tumors that develop from the protective covering of nerves. These growths can occur throughout the body and may lead to issues such as pressure symptoms and aesthetic or functional anomalies, depending on their specific location. A less common type, known as ancient schwannomas, grows slowly and is marked by cystic and necrotic deterioration within the tumor tissue. It is exceptionally rare for these tumors to appear in the spinal canal, especially within the sacral region. Often, these tumors are discovered by accident or through the manifestation of pain and neurological symptoms like lower back discomfort, loss of sensation, or tingling. In the case discussed here, the patient, a farmer by profession, experienced no notable symptoms apart from a sensation of abdominal fullness after eating, persisting for a month. Upon seeking medical advice, an abdominal ultrasound was conducted, revealing a mass. Further investigation through CT identified the mass as originating from the second sacral vertebra, and a histological evaluation confirmed the diagnosis of ancient schwannoma. **Introduction:**Schwannomas, or neurilemmomas, are non-malignant growths originating from the embryonic neural crest cells that form the nerve sheath [1]. Typically manifesting in individuals during their forties and fifties, there's a slight inclination towards female patients [2]. These growths can appear anywhere within the neural system but are predominantly found on the head, the inner sides of limbs, and the torso. While schwannomas are somewhat common in the cervical and lumbar regions of the spine, their occurrence in the sacral spine is notably rare [3]. The symptoms vary based on the tumor's location, with back discomfort and radicular signs being the most common [4]. Ancient schwannomas differ from their more common counterparts by exhibiting several degenerative changes, such as calcification, cyst development, hemorrhage, and hyalinization around blood vessels [5]. These tumors do not possess unique clinical or imaging characteristics, making histological analysis crucial for accurate diagnosis [6]. This case report highlights a spinal mass that posed diagnostic challenges and was ultimately identified as an ancient schwannoma through surgical removal and subsequent histopathological evaluation [7].

CASE REPORT

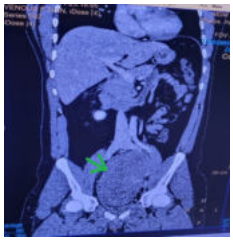
A 29-year-old male farmer experienced persistent abdominal fullness, particularly after meals, over the past month. Despite his ongoing engagement in daily activities, a physical examination revealed his abdomen to be swollen but not painful upon touch. Initial diagnostic ultrasound of the abdomen disclosed a large, irregularly shaped lesion with a dumbbell appearance, measuring 15.8 x 6.8 x 7.7 cm. Further detailed assessment using CT scans of the entire abdomen identified a distinct, lobulated, low-density mass approximately 12 x 8.9 x 13.5 cm in size. This mass exhibited uneven enhancement following contrast administration, situated in the pelvic region, specifically within the pre and retrorectal extraperitoneal space, alongside adjacent inflammation of the fat tissue [8].

The mass extended into the left second sacral (S2) foramina, causing it to widen and the surrounding bone to become sclerotic, suggesting its origination from this area. It exerted pressure on the urinary bladder and the junction between the rectum and sigmoid colon from the front, yet fat planes were preserved, indicating separation from these structures [9].

From behind, the mass pressed against the rectum, also maintaining fat planes, which suggests no direct invasion into the intestines or visible signs of intestinal blockage. The mass was in close proximity to the space before the vertebral bodies from L5 to S3 without eroding the bone. On the left postero-lateral side, it was in direct contact with the piriformis muscle, showing a loss of the separating fat layer. Laterally, it displaced the common and external iliac arteries, though these vessels remained functionally unimpeded, as indicated by normal contrast flow [10].



Axial CT image showing tumor



CT showing tumour

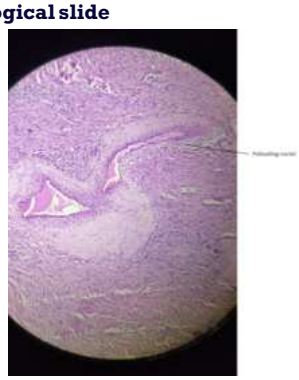
The lesion was identified to receive its blood supply from the left common iliac artery, indicating a significant vascular connection. Additionally, a linear, low-density filling defect was observed in the left external iliac vein, suggesting a partial thrombosis. This condition extended to the left common and superficial femoral vein, where collateral

formations were noted around the bladder and adjacent areas, further indicating vascular complications associated with the mass. Despite these findings, there was no indication of widening in the neural foramina nor any sign of bone erosion, pointing to the localized nature of the lesion without extensive skeletal involvement [11].

Given the lesion's characteristics and potential origins, the differential diagnosis included a peripheral nerve sheath tumor, sarcoma, or leiomyosarcoma, each suggesting varying degrees of malignancy and origins related to nerve or muscle tissue. The decision to proceed with surgical removal was based on the encapsulated and cystic nature of the tumor, aiming for an en bloc resection to minimize the risk of recurrence and ensure complete removal [12].

The surgery was performed successfully without any intraoperative complications, showcasing the medical team's expertise in handling complex pelvic masses. The patient's recovery was notably swift and complication-free, allowing for his discharge a few days post-operation. This outcome not only highlights the effectiveness of the surgical approach but also the importance of early detection and intervention in managing potentially life-altering pelvic tumors [13].

Histopathological slide



DISCUSSION

Schwannomas, benign tumors deriving from Schwann cells that insulate nerve fibers, are pivotal in the understanding of peripheral nerve pathologies. These tumors are characterized by their slow growth and a propensity to remain asymptomatic for extended periods, often leading to delayed diagnosis. Schwannomas are encapsulated, thereby facilitating surgical resection, a preferred method for managing symptomatic cases. Histologically, schwannomas are distinguished by two cellular patterns: Antoni A and Antoni B areas. Antoni A areas are densely cellular, showcasing spindle cells with palisading nuclei, while Antoni B areas are less cellular, with a myxoid [14].

Ancient schwannomas represent a unique subclass, accounting for less than 1% of all schwannomas [15]. These tumors exhibit pronounced degenerative changes, including cystic necrosis, calcification, and hemorrhage, attributed to their prolonged growth period. Such features can obscure the typical histological architecture of schwannomas, complicating the diagnosis. The term "ancient" reflects these extensive degenerative changes rather than the age of the tumor itself [16].

Clinically, the presentation of schwannomas varies significantly depending on their location. While often asymptomatic, large tumors can exert pressure on adjacent structures, leading to pain, neurological deficits, or organ dysfunction, as seen in our case with abdominal fullness and potential urinary effects due to bladder compression. The rarity of sacral schwannomas, especially ancient variants, adds a layer of complexity to clinical and radiological diagnosis, underscoring the need for a high index of suspicion [17].

Diagnostic imaging, including MRI and CT scans, plays a crucial role in the initial identification and localization of these tumors. However, definitive diagnosis relies on histopathological examination, as imaging cannot conclusively differentiate between schwannomas and other similar neoplasms like neurofibromas or malignant peripheral nerve sheath tumors [18].

The management of schwannomas generally involves surgical resection, aiming for complete removal to prevent recurrence and alleviate symptoms. In the case of ancient schwannomas, surgery may be challenging due to the tumor's size and involvement with surrounding structures but remains the treatment of choice. The surgical approach should be meticulously planned to minimize damage to adjacent nerves and tissues [19]

CONCLUSION

The presentation of an ancient schwannoma in the sacral spine, as delineated in this case, underscores a rare but significant clinical entity within the domain of spinal tumors. Ancient schwannomas are distinguished from their more common counterparts by their pronounced degenerative changes and slow growth rate, characteristics that contribute to their elusive nature in clinical and radiological diagnostics. The case highlights the importance of a multidisciplinary approach in the diagnosis and management of such tumors, given their potential to mimic other neoplasms and the complex surgical considerations they entail.

The rarity of sacral ancient schwannomas poses a considerable challenge to healthcare providers, necessitating heightened awareness and suspicion, especially in patients presenting with nonspecific symptoms like abdominal fullness or back pain. This report emphasizes the critical role of comprehensive imaging and histopathological examination in establishing a definitive diagnosis, which in turn guides therapeutic decision-making.

Surgical resection remains the cornerstone of management for ancient schwannomas, aimed at alleviating symptoms and preventing potential complications. The successful outcome in this case, marked by the patient's uneventful recovery and discharge, reaffirms the efficacy of surgical intervention, even in the context of large and anatomically complex tumors. It also highlights the necessity for ongoing research and reporting on such cases to enhance our understanding of ancient schwannomas, improve diagnostic accuracy, and refine surgical techniques to optimize patient outcomes.

In conclusion, this case report contributes to the growing body of literature on ancient schwannomas, providing valuable insights into their presentation, diagnosis, and management. It underscores the need for continued vigilance and a comprehensive diagnostic workup in cases of unusual spinal tumors to ensure timely and effective treatment. Future studies are encouraged to further elucidate the biological behavior of ancient schwannomas and to explore advances in surgical and non-surgical therapies, thereby improving the quality of care for patients afflicted with this rare condition.

Conflicts of interest

The authors declare no conflicts of interest.

Footnotes

This study was carried out at the Mathura Das Mathur Hospital, Jodhpur, Rajasthan, India.

Ethics Statement: The authors retain informed consent signed by the patient for authorizing the data publication and the manuscript is as by the Institutional Ethics Committee rule.

Financial support: The authors declare no financial support was received.

REFERENCES

1. Nadeem M, Mansoor S, Assad S, Ilyas F, Qavi AH, Saadat S, et al. Spinal schwannoma with intradural intramedullary hemorrhage. *Cureus.* 2017;9:e1082. [PMC free article] [PubMed] [Google Scholar]
2. Drevelagas K, Bangeas P. Intradural extramedullary schwannoma of thr thoracic spine with intratumoral hemorrhage. *OMICS J Radiol.* 2017;6:272. [Google Scholar]
3. Hdeib A, Goodwin CR, Sciubba D, Bydon A, Wolinsky JP, Witham T, et al. Hemorrhagic thoracic schwannoma presenting with intradural hematoma and acute paraplegia after spinal manipulation therapy. *Int J Spine Surg.* 2016;10:42. [PMC free article] [PubMed] [Google Scholar]
4. Ciappetta P, D'Urso PI, Colamaria A. Giant craniovertebral junction hemorrhagic schwannoma: Case report. *Neurosurgery.* 2008;62:E1166. [PubMed] [Google Scholar]
5. Conti P, Pansini G, Mouchaty H, Capuano C, Conti R. Spinal neurinomas: Retrospective analysis and long-term outcome of 179 consecutively operated cases and review of the literature. *Surg Neurol.* 2004;61:34–43. [PubMed] [Google Scholar]
6. Parmar H, Patkar D, Gadani S, Shah J. Cystic lumbar nerve sheath tumours: MRI features in five patients. *Australas Radiol.* 2001;45:123–7. [PubMed] [Google Scholar]
7. Yeh HM, Leung JH, Huang KC, Tung CL, Huang CL, Huang KM. A long segmental hemorrhagic spinal schwannoma with atypical presentation. *J Radiol Sci.* 2011;36:191–4. [Google Scholar]
8. Sahoo RK, Das PB, Sarangi GS, Mohanty S. Acute hemorrhage within intradural extramedullary schwannoma in cervical spine presenting with quadriparesis. *J Craniovertebr Junction Spine.* 2015;6:83–5. [PMC free article] [PubMed] [Google Scholar]
9. Zhang HZ, Li Y, Han Y, Wang X, She L, Yan Z, et al. Spontaneous acute hemorrhage of intraspinal canal cellular schwannoma with paraplegia: A case report. *Br J Neurosurg.* 2015;29:425–7. [PubMed] [Google Scholar]
10. Vaibhav N, Mahalingam SS, Parthiban JK. Hemorrhage within the schwannoma of thoracic spinal cord presenting as acute rapidly progressive paraplegia: A rare case. *J Spinal Surg.* 2016;3:160–2. [Google Scholar]
11. Jaiswal A, Shetty AP, Rajasekaran S. Giant cystic intradural schwannoma in the lumbosacral region: A case report. *J Orthop Surg (Hong Kong)* 2008;16:102–6. [PubMed] [Google Scholar]
12. Prasad GL, Kongwad LI, Valiathan MG. Spinal intradural schwannoma with acute intratumoural haemorrhage: Case report and review. *J Clin Diagn Res.* 2016;10:PD01–3. [PMC free article] [PubMed] [Google Scholar]
13. Vázquez-Barquero A, Pascual J, Quintana F, Figols J, Izquierdo JM. Cervical schwannoma presenting as a spinal subdural haematoma. *Br J Neurosurg.* 1994;8:739–41. [PubMed] [Google Scholar]
14. Jenkins AL, 3rd, Ahuja A, Oliff AH, Sobotka S. Spinal schwannoma presenting due to torsion and hemorrhage: Case report and review of literature. *Spine J.* 2015;15:e1–4. [PubMed] [Google Scholar]
15. Tanaka H, Kondo E, Kawato H, Kikukawa T, Ishihara A, Toyoda N. Spinal intradural hemorrhage due to a neurinoma in an early puerperal woman. *Clinical Neurology and Neurosurgery.* 2002;104:303–5. [PubMed] [Google Scholar]
16. Cohen ZR, Knoller N, Hadani M, Davidson B, Nass D, Ram Z. Traumatic intratumoral hemorrhage as the presenting symptom of a spinal neurinoma. Case report. *Journal of Neurosurgery.* 2000;93(2 suppl):327–9. [PubMed] [Google Scholar]
17. Uemura K, Matsumura A, Kobayashi E, Tomono Y, Nose T. CT and MR presentation of acute hemorrhage in a spinal schwannoma. *Surgical Neurology.* 1998;50:219–20. [PubMed] [Google Scholar]
18. Ichinose T, Takami T, Yamamoto N, Tsuyuguchi N, Ohata K. Intratumoral hemorrhage of spinal schwannoma of the cauda equina manifesting as acute paraparesis—case report. *Neurologia medico-chirurgica (Tokyo)* 2009;49:255–7. [PubMed] [Google Scholar]
19. Ng PY. Schwannoma of the cervical spine presenting with acute hemorrhage. *Journal of clinical neuroscience.* 2001;8:277–8. [PubMed] [Google Scholar]
20. Smith RA. Spinal subdural hematoma, neurilemmoma, and actute transverse myelopathy. *Surgical Neurology.* 1985;23:367–70. [PubMed] [Google Scholar]