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Gigantic retroperitoneal pararenal ancient schwannoma masquerading as a mesenchymal malignancy: A diagnostic conundrum

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ABSTRACT

Ancient schwannoma is a rare benign variant of schwannoma with marked degenerative changes, often mimicking malignancies, particularly when retroperitoneal and pararenal. A 34-year-old woman presented with a rapidly growing 15 cm retroperitoneal pararenal mass. Imaging suggested an aggressive malignancy. Surgical resection and histopathology revealed a well-encapsulated tumor with degenerative changes, nuclear atypia, and strong S100 positivity, confirming ancient schwannoma. Complete excision achieved negative margins, and follow-up showed no recurrence. Diagnostic challenges arise from its rarity and atypical features. Histopathology is crucial for differentiation from malignancies. Ancient schwannomas should be considered for large retroperitoneal masses to ensure accurate diagnosis and management.

1. Introduction

Schwannomas are typically well-circumscribed, encapsulated, benign peripheral nervous system tumors, arising from Schwann cells in the nerve sheath. These tumors exhibit a very low risk of malignancy and can occur across all age groups, with the highest incidence in people aged 30–60. Ancient schwannoma, a rare and benign histopathological variant, is distinguished by a long duration and degenerative changes within the tumour, often affecting older adults.[1]

While schwannomas are usually small (1–2 cm), they can uncommonly grow larger, which, along with the presence of long-standing degenerative changes, can very well mimic a malignant neoplastic etiology. Moreover, retroperitoneal humungous mass lesions are frequently aggressive mesenchymal malignancies like dedifferentiated liposarcoma, leiomyosarcoma or undifferentiated pleomorphic sarcoma (UPS). Pararenal retroperitoneal schwannomas are exceptionally rare, with only a handful of cases reported in international literature. Herein, we elucidate the complete clinical, radiological and pathological spectrum of the present case of a gigantic pararenal retroperitoneal ancient schwannoma in a 34-year-old woman masquerading as a sarcoma. This case study describes the atypical presentation of a benign entity owing to its enormous size, occurrence at an unusual site, and exhibiting marked degenerative atypia which can pose diagnostic and management dilemmas to urologists, uroradiologists and uropathologists alike. However, arriving at an accurate diagnosis remains pertinent as the prognostic outcome is entirely different and timely diagnosis can help guide clinicians in formation of appropriate management and follow-up plans.

2. Case presentation

A 34-year-old woman presented with a progressively enlarging lump in the right lumbar region, which had grown from 4 × 4 cm to 15 × 14 cm over a span of three months. She reported intermittent pain that resolved spontaneously, with no other abdominal symptoms. On examination, the lump had a dull percussion note, was non-pulsatile, and was markedly tender on deep palpation.

Contrast-enhanced MRI (CE-MRI) revealed a large, solid-cystic mass lesion in the right anterior pararenal space measuring 15.2 cm (antero-posteriorly) x 14.7 cm (transversally) x 18.1 cm (cranio-caudally) with cystic changes, areas of haemorrhage, and fluid levels, creating a mass effect on the adjacent anatomical organs (Fig. 1A–C). Contrast-enhanced Computed Tomography (CECT) scan exhibited the presence of a large hypodense lesion with mild enhancement and small amorphous calcification in the right pararenal lumbar region which was displacing the right colon antero-medially, indenting the anterior surface of right kidney, abutting the inferior surface of the right lobe of the liver and the infero-medial surface of gall bladder with inferior vena cava pushed along the medial border of the lesion (Fig. 1D). Radiological findings suggested a primary retroperitoneal tumor, possibly arising from the

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right pararenal space. Radiological differential diagnoses included a large mesenchymal malignancy with degenerative cystic changes with possible differentials of a pleomorphic liposarcoma or UPS.

The patient underwent an exploratory laparotomy and the entire mass lesion was resected with adequate margins and sparing the kidney. The excised retroperitoneal mass was sent for histopathological evaluation in 10 % neutral buffered formalin. Gross examination showed a well-encapsulated, grey-brown soft tissue mass measuring 18 × 15 × 14 cm. Serosanguinous fluid oozed out on serial sectioning, with variegated solid and multiloculated cystic areas varying in diameter from 1 to 12 cm in diameter (Fig. 2A). Extensive representative sampling was done to assess the heterogeneity of the neoplasm adequately. Routine 5-μm thick sections were cut from the paraffin-embedded and formalin-fixed tissue and were subsequently stained with Hematoxylin and Eosin (H&E) stains.

Multiple microscopic sections examined revealed a well-encapsulated, circumscribed spindle cell neoplasm with variable alternating areas of hypercellularity and hypocellular myxoid zones (Fig. 2B), cystic degeneration, haemorrhage, and marked nuclear atypia. Markedly large areas of myxoid degeneration were evident (Fig. 2C). Individual tumor cells were spindle to elongated with a moderate amount of eosinophilic cytoplasm, and indistinct cellular membranes. They had elongated nuclei exhibiting pleomorphism with irregular nuclear contours (Fig. 2D). However, the chromatin was bland and homogenous with inconspicuous nucleoli. Notable features included infarction, haemorrhage (Fig. 3A), hyalinization, calcification (Fig. 3B), clusters of foamy macrophages (Fig. 3C), and the occasional presence of Verocay bodies (Fig. 3D). There was no evidence of tumor necrosis and mitotic activity was sparse. A comprehensive immunohistochemistry (IHC) panel of antibodies was applied on 2-μm thick sections on Poly-L-Lysine coated slides to clinch an accurate diagnosis. On IHC, the tumor cells showed immunopositivity for S100 (Fig. 4A), Calretinin (Fig. 4B), CD56 (Fig. 4C), and Vimentin, and were negative for neurofilament, synaptophysin, and chromogranin, NSE, MDM2, CDK 4, CD 68, Desmin, Myogenin, Smooth Muscle Actin (SMA), and CD 34. Ki67 proliferation index was very low, around 1–2% (Fig. 4D). Based on the concordant histomorphological and immunohistochemical findings a final diagnosis of ancient schwannoma in the right pararenal region was rendered.

The resected surgical tumor margins were negative and the patient was hemodynamically stable in the post-operative period. The patient has been kept on follow-up in the surgical outpatient department and has shown no signs of recurrence at the site of excision or elsewhere in the body.

3. Discussion

Schwannomas are slow-growing, solitary, well-encapsulated, and often asymptomatic benign tumors of neurogenic origin. They typically arise from the Schwann cells of the peripheral nerve sheath of neurons. Schwannomas exhibit a definite predilection for the head and neck region and the extremities and commonly involve the sensory nerves as compared to the motor nerves. Although they usually present with the median range of 3–5 cm in its largest dimension, they can sometimes grow to enormous sizes, creating diagnostic dilemmas for clinicians and radiologists. When presenting as such gigantic neoplasms, they can produce symptoms due to pressure or mass effect on surrounding structures. Not only in diagnosis, they can further add to the operative difficulty in adequate resection of the tumor, and at the same time, preserving all the vital adjacent structures.[2,3]

Schwannomas are classified into various histological subtypes, including ancient, microcystic-reticular, epithelioid, cellular, psammomatous, and melanotic schwannomas. The ancient variant, is a rare benign form, which was first described by Ackermann and Taylor in 1951. Tumors which grow to become exceedingly large neoplasms, can have deficit vascular supply within the tumor proper, creating hypoxic environment for the tumor, due to which marked degenerative changes can be evident including cystic change, hyalinization, haemorrhage, accumulation of cyst or hemosiderin-laden macrophages, infarction, calcification, and myxoid degeneration. Moreover, classical Antoni A (hypercellular region with presence of Verocay bodies) and Antoni B

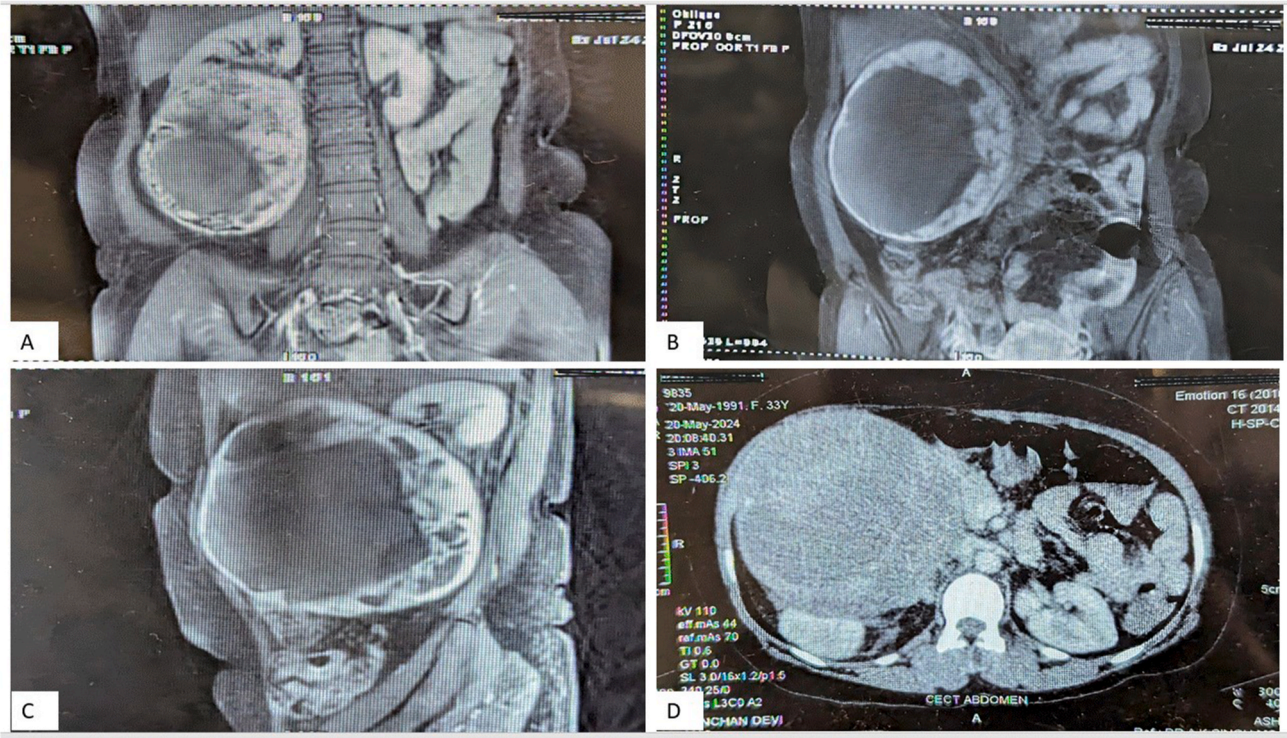

Fig. 1. Radiological findings: CE-MRI (Figures A–C) and CECT (Figure D) revealing the presence of a large well defined, rounded, solid cystic mass lesion seen epicentered in the right lumbar region in the anterior pararenal space.

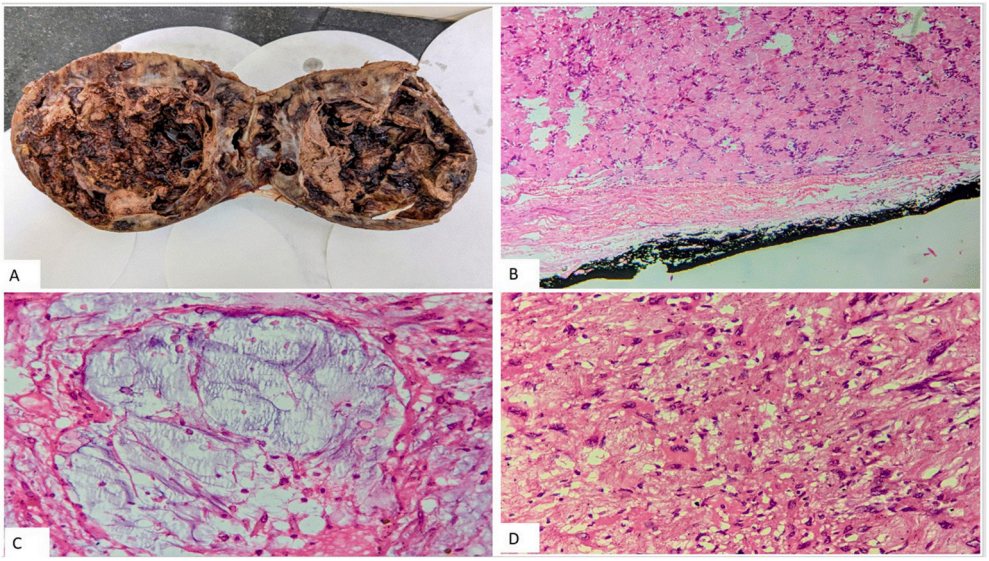

Fig. 2. – Pathological findings: Figure A – Gross findings: A well-encapsulated grey-brown globular soft tissue mass with solid cystic and hemorrhagic areas. **Figures B – D) – Microscopic findings:** (B) - Multiple sections exhibiting the presence of a well-encapsulated tumor (Hematoxylin and eosin x100), (C) - Areas of myxoid degeneration (Hematoxylin and eosin x400), (D) - Tumor cells exhibiting degenerative atypia with absent mitotic activity. (Hematoxylin and eosin x400).

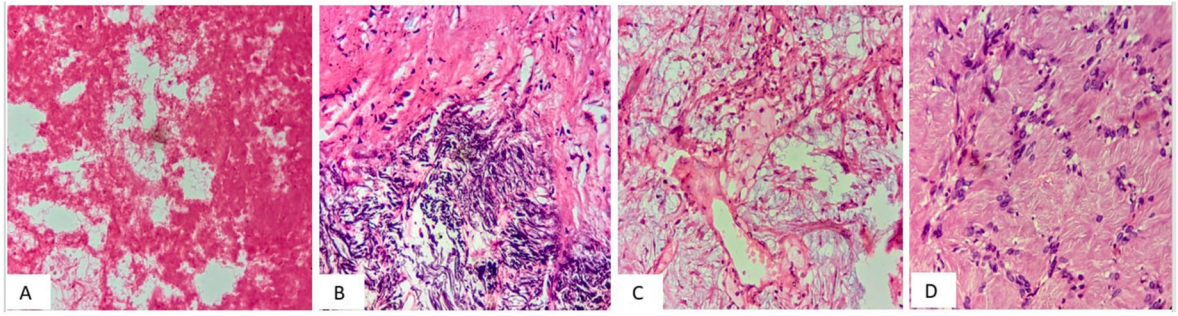

Fig. 3. Pathological findings: Sections exhibiting (A) - areas of haemorrhage, (B) - foci of calcification, (C) - cluster of foamy macrophages, and (D) - presence of occasional Verocay bodies. (Hematoxylin and eosin x400).

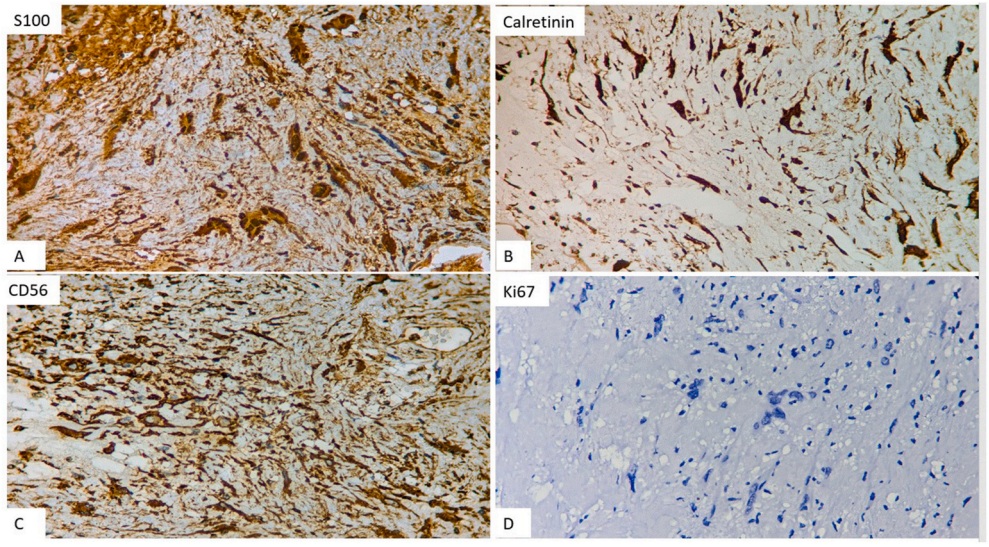

Fig. 4. – Immunohistochemical findings: Tumor cells exhibiting positivity for (A) - S100, (B) - Calretinin, (C) - CD56, with a (D) - low Ki 67 proliferation index. (Immunohistochemistry, x400).

(hypocellular region) which are typically seen in schwannomas may not be appreciated in ancient schwannomas. Also, marked degenerative atypia can be striking in the spindle cells which can lead to the suspicion of a malignant neoplasm. Therefore, careful histopathological evaluation remains crucial in arriving at an accurate diagnosis. Absence or sparse mitoses despite the presence of nuclear atypia is an extremely helpful clue which points towards a benign etiology with degenerative changes.[4,5]

Furthermore, IHC plays a pivotal role in ruling out differentials and clinching a definitive diagnosis. As in this case diffuse and intense positivity for S 100, suggested a neural origin of the neoplasm. Malignant peripheral nerve sheath tumor (MPNST), which is a malignant counterpart and exhibits marked pleomorphism, on the other hand shows weak, patchy or negative immunostaining for neural markers like S 100. Moreover, an extremely low Ki 67 labeling index (i.e. 1-2%) further favoured a benign etiology. Another close differential included Ganglioneuroma, which typically occurs in the retroperitoneal region along with other sites such as the neck, posterior mediastinum, pelvis, and adrenal glands owing to the anatomical presence of sympathetic system neurons at these locations. However, histomorphological features like absence ganglion cells admixed within schwannian stroma and negative immunoexpression for synaptophysin, NSE, and neurofilament further helped in ruling out its possibility. Retroperitoneal mass lesions, especially those presenting as large lesions, frequently arouse the possibility of aggressive mesenchymal malignancies which were meticulously ruled out by histomorphological evaluation and application of an extensive IHC panel. A negative immunohistochemical expression for MDM 2 and CDK 4 negated the possibility of a de-differentiated liposarcoma, negative SMA, Desmin, and Myogenin helped in ruling out leiomyosarcoma and spindle-cell rhabdomyosarcoma, and negative CD 34 ruled out fibroblastic origin as in solitary fibrous tumor. Therefore, the corroborative histopathological and immunohistochemical features helped arrive at a conclusive diagnosis of a retroperitoneal ancient schwannoma.

Retroperitoneal schwannomas are uncommon, representing just 1–3% of all schwannomas and a mere 1 % of all retroperitoneal neoplasms. Imaging characteristics that raise suspicion for ancient schwannoma include a hypervascular soft-tissue mass with cystic changes on MRI, alongside amorphous calcifications on radiography. A fludeoxyglucose-18 (FDG) positron emission tomography (PET) scan can show a high level of FDG uptake in schwannomas, a benign nerve sheath tumor. This can make it difficult to distinguish schwannomas from malignant tumors before a biopsy or surgery. Moreover, a small biopsy may not be representative due to the presence of marked degenerative changes and the heterogeneity of the large tumor. Therefore, histopathological examination of the excised mass with ancillary studies of immunohistochemistry remains the cornerstone for establishing a definite diagnosis. Key histological features of ancient schwannoma include cystic degeneration, haemorrhage, calcification, perivascular hyalinization, macrophages or siderophages, myxoid change, xanthomatous change, and nuclear atypia—all observed in our case. Given the size and nuclear atypia in these tumors, a low mitotic count and low Ki 67 index is essential for excluding malignancy, as highlighted in the present study.[6–8] Table 1 enlists and describes the previously reported pararenal retroperitoneal ancient schwannomas.

Diagnosing large and humungous retroperitoneal ancient schwannomas can be challenging owing to their rarity, atypical location, and lack of specific clinical and radiological presentation. Complete surgical excision with negative surgical margins remains the preferred treatment, as it aims to preserve neurological function and minimize complications.

Ancient schwannomas have an excellent prognostic outcome. Complete surgical excision is typically curative, although incomplete resection may subsequently lead to recurrence.

Table 1
Summary of pararenal schwannoma cases previously reported in the literature.

S. no	Authors	Age	Sex	Location	Size	Histopathology
1	Miyagi T et al.[9]	64	male	Left pararenal space	3.5 × 2.0cm	Schwannoma
2	Patrinou A et al.[10]	24	female	Left pararenal space	9.4 × 4 × 4cm	Schwannoma
3	Peng X et al.[11]	35	female	left pararenal space	13.x8.5 × 6.5cm	Schwannoma
4	Present case	34	female	Right Pararenal space	17.5 × 15 × 12cm	Ancient schwannoma

4. Conclusion

The present study sheds light on the challenges of accurately diagnosing an enormously large retroperitoneal neoplasm, initially presumed to be of malignant mesenchymal etiology. It emphasizes that despite of their rare occurrence and atypical clinico-radiological presentation, ancient schwannomas should be kept as a plausible differential diagnosis when coming across large retroperitoneal mass lesions. It also reiterates the pivotal role of histopathology for a definitive diagnosis, prognostication and formation of appropriate management and follow-up plans for the patients.

CRediT authorship contribution statement

Neha Aggarwal: Writing – review & editing, Writing – original draft, Visualization, Validation, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Shaivy Malik:** Writing – review & editing, Writing – original draft, Visualization, Validation, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Charanjeet Ahluwalia:** Writing – review & editing, Visualization, Validation, Supervision, Resources, Project administration, Methodology.

Statement of ethics

Written informed consent was taken from the patient to participate in this study and for the publication of any potentially identifiable images or data included in this article. Ethical review and approval are not required and are provided a waiver for the publication of case reports as per institutional requirements.

Data availability statement

No new data was generated during this study.

Declaration of generative AI in scientific writing

No generative AI help was used in scientific writing of this manuscript.

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Conflict of interest statement

The authors declare no conflict or competing interests.

The authors declare that the study was conducted in the absence of any commercial or financial relationships that could be interpreted as a potential conflict of interest. No funding was received for the conduct of

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