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A Rare Case Report of Retroperitoneal Ancient Schwannoma.

Fathima Nifra M*, Hemalathaganapathy, and Parijatham BO.

Department of Pathology, Sree Balaji Medical College and Hospital, Bharath University, Chrompet, Chennai – 600044, Tamil Nadu, India.

ABSTRACT

Schwannoma or neurilemmoma is a well-defined tumor arising from the nerve sheath (schwann cells). It is most commonly seen in the head, neck and extremities, rarely arises in the retroperitoneum. Ancient variety of schwannoma is a long standing tumour seen in the middle aged groups with degenerative changes. Schwannomas are difficult to diagnose using imaging only so tissue sampling through CT guided biopsy is essential to confirm the diagnosis before surgery. Immunostaining revealed S 100 positivity and histopathology confirmed the diagnosis. Herein we report a rare case of benign retroperitoneal paravertebral ancient schwannoma.

keywords: Ancient schwannoma, Retroperitoneal schwannoma, Schwannoma

**Corresponding author*

CASE REPORT

A 44 year old male presented to our hospital with history of fever and abdominal pain. He underwent a CT scan of the abdomen and pelvis i reported a 3.5x 3.7 cm retroperitoneal mass in left paracolic gutter mass at level of L2 L3. CT guided biopsy revealed a 4x5 cm retroperitoneal mass at site of left paracolic gutter behind the descending colon adjacent to the aorta. The patient underwent surgical excision of the mass

Gross examination revealed a soft tissue mass measuring 6x5x3 cm with irregular and friable cut surface. External surface appeared nodular and well circumscribed.

Histopathological examination revealed schwannoma showing collection of foamy histiocytes and areas of hyalinization. Verocay bodies with Antoni A (highly ordered cellular component, well -organised spindle cells in palisading pattern) and Antoni B (less cellular, loose textured pleomorphic cells against a myxoid background) along with areas of thrombosed and hyalinised blood vessels were present focally. Degenerative changes like cystic degeneration and foci of calcification were seen. Diagnosis was confirmed with immunohistochemistry, which were diffusely positive for S-100 protein.

DISCUSSION

Schwannomas are benign, slow growing tumors that originate from Schwann cells. They seldom cause symptoms and are often discovered incidentally. The common sites of involvement include the head and neck, the flexor surfaces of the upper and lower extremities, the posterior mediastinum in the thorax and on the trunk [1,2]. Retroperitoneal schwannomas are rare tumours, with incidence of 0.7% to 2.6% of all schwannomas. They occur most commonly between 40 and 60 years of age, with a male/female ratio of 2:3. Most schwannomas are benign, although malignant cases are rarely known to occur. Malignant transformation [neurofibrosarcoma] is usually observed in cases with underlying Von Recklinghausen's disease [3, 4]. Gross appearance, of schwannomas are usually solitary, well circumscribed, firm, smooth-surfaced tumors. [4]. Histologically they are made up of elongated bipolar spindle cells with a focal nuclear palisading pattern. There are areas of high cellularity named Antoni A and with low cellularity and myxoid matrix named Antoni B [5]. "Ancient" schwannoma, was initially mentioned by Ackerman and Taylor as a degenerative change occurring in a long standing schwannoma, was characterised by nuclear hyperchromasia, mild nuclear pleomorphism, stromal oedema, fibrosis and xanthomatous changes [6]. Degeneration is due to central tumor necrosis as the schwannoma grows to a size beyond the capacity of its blood supply [7]. Schwannomas are characterized immunohistochemically by S-100 positivity [8].

CONCLUSION

MRI is the imaging modality of choice in demonstrating tissue heterogeneity and anatomic location of the tumour. However, needle biopsy should be regarded as the diagnostic gold standard, as none of the currently available imaging modalities provide sufficient information to exclude malignancy. Wide surgical removal is the treatment of choice and malignant transformation is rare. This case is presented for its rarity.



Figure 1



Figure 2: Gross image showing external surface nodular and well circumscribed.

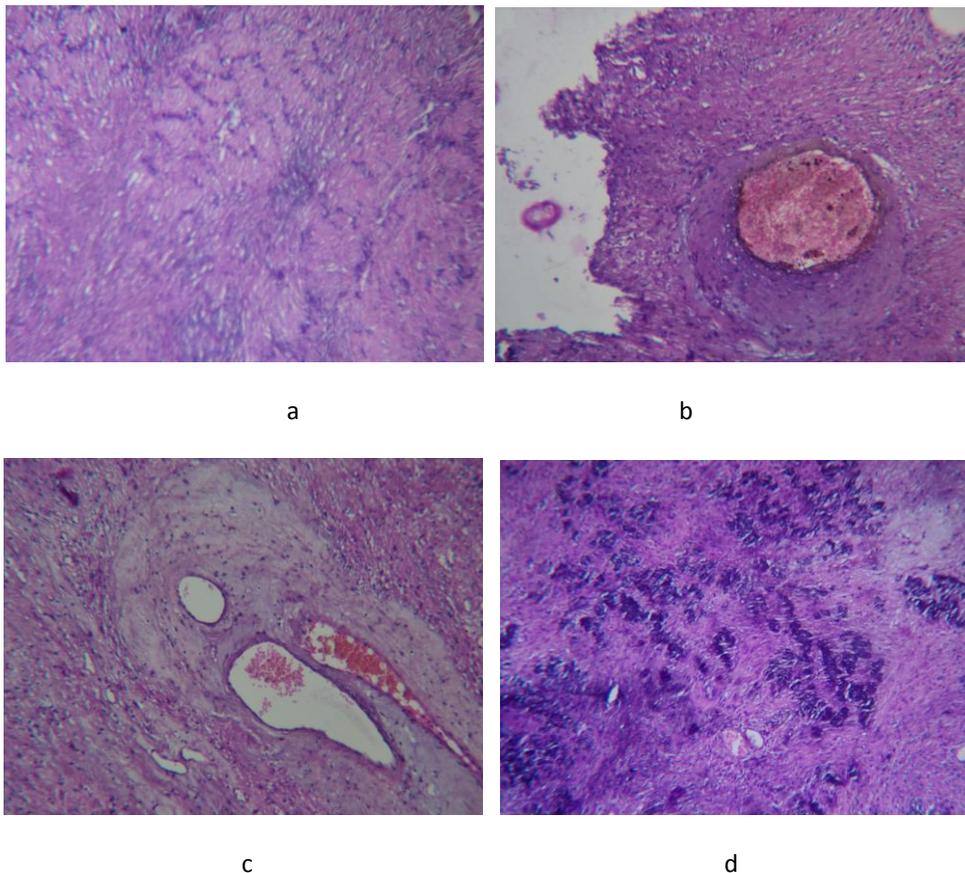


Figure 3: Soft tissue measuring 6x5x3 cm with irregular and friable cut surface.

Histopathological images of the specimen (a) 10x image showing high cellularity Antoni A regions , low cellularity Antoni B regions.(b) 20x image showing thrombosed blood vessel.(c) 20 x image showing hyalinised blood vessel.(d)10 x image showing foci of calcification.

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