Benign schwannoma of the abdominal wall: A rare case report and a review of literature

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ABSTRACT

Tumors of the peripheral nerve sheath (neurilemmomas or schwannomas) are uncommon, affecting approximately 5% of adults. The incidence of such a tumour over the abdominal wall is extremely rare. They are mostly benign and require complete surgical excision to prevent a sequel. Here, we report a single case of a 22-year-old male presenting with a lump over the left flank and was treated in our teaching medical institution in Eastern India. Various imaging modalities (ultrasonography and magnetic resonance imaging) were used to diagnose this tumor, before it was surgically excised. Histopathology confirmed the swelling to be a benign schwannoma of the abdominal wall. We have also reviewed the literature associated with this rare clinical entity. To the best of our knowledge, only a handful of case reports on abdominal wall schwannomas have been reported so far.

Key words: *Antoni A, Antoni B, Nerve sheath, Neurilemmoma, Schwannoma, Verocay bodies*

Peripheral nerve sheath tumors include a myriad of benign and malignant entities, and most of these neoplasms show Schwann cell differentiation. Three neoplasms, namely schwannoma, neurofibroma, and malignant peripheral nerve sheath tumor, are the most common to occur. Among them, neurilemmomas or schwannomas are rare ectodermal tumors of the peripheral nerve sheath that arises from the Schwann cells [1]. They are commonly seen in the flexor aspect of the extremities, trunk, head, and neck. Abnormal locations include the retroperitoneum, pelvis [2], perineum [3], and inguinal canal mimicking the inguinal hernias [4]. Histologically, schwannomas are distinguished by the presence of areas of high and low cellularity, called Antoni A and B respectively, and they stain positively with S100 during immunohistochemistry. Treatment of schwannoma is complete excision, but they are notorious for recurrence, after an incomplete removal.

The literature associated with schwannomas *per se* is diverse, and various authors have described their personal experiences in dealing with such tumors in various abnormal locations. A benign schwannoma of the abdominal wall is a rare clinical entity. To the best of our knowledge, only a handful of case reports that describe the nature and behavior of this tumor have been reported so far. We report a single case of a benign schwannoma in a young male from a teaching medical institution in West Bengal, India.

CASE REPORT

A 22-year-old male presented to the surgical outpatient department with a gradually progressing swelling over the left flank. The mass had increased to its present size of approximately 5 cm over the past 1 year. The patient was apparently symptomless without any history of weight loss, anorexia, trauma, or paraesthesia. There was no family history of such lumps.

On examination, his vitals and general survey were within normal limits. His abdomen had a well-defined, firm, 5 cm swelling over the left flank. The swelling was slightly tender on palpation, not fixed to the skin or deeper structures, but it became less prominent on contracting the muscles of the abdominal wall (Fig. 1). All routine blood parameters were within normal limits.

On ultrasonography (USG), a well-defined, 6.2 cm×4 cm, encapsulated, ovoid, heterogeneously hypoechoic space-occupying lesion was noted within the muscle plane indenting the underlying structures. The lesion also showed posterior acoustic enhancement, had hypoechoic extensions on both sides of the lesion (peripheral nerve continuity) (Figs. 2a and b). On color Doppler, internal vascularity was demonstrated. Rest of the abdomen was within normal limits. Magnetic resonance imaging (MRI) was done, and it revealed a well-defined, round-to-ovoid 6 cm×3.7 cm×3.2 cm space-occupying lesion between the external and internal oblique muscles indenting the deeper structures on the left anterolateral abdominal wall. On T1-weighted imaging (T1WI), it was iso to hypointense (Fig. 3a). On T2-weighted (T2WI) image, it was heterogeneously hyperintense (Fig. 3b and c). On fat suppression sequence (inversion recovery), it was hyperintense without any surrounding inflammatory changes (Fig. 3d) and avidly enhanced with contrast study.

Imaging features were consistent with a peripheral nerve sheath tumor. A decision for an *en bloc* surgical excision was taken. After

administration of spinal anesthesia, the swelling was approached through the left flank after dividing the external oblique muscle (Fig. 4a). The tumor was seen lying in between the external and internal oblique muscles, and it extended superiorly to the lower border of the ribs. A combination of blunt and sharp dissection was used to dissect the swelling. A small nerve was seen to enter and exit the swelling (Fig.4b). The tumor was completely excised and sent for histopathology.

On macroscopic examination, the swelling was pale yellow (Fig. 5a and 5b). The microscopic section showed an admixture of hypercellular areas (Antoni A) with dense eosinophilic spindle cells arranged in fascicles and hypocellular areas where the spindle cells are spread apart by a prominent extracellular matrix (Antoni B) (Fig. 6a). The Antoni A areas with the tumor cells have nuclei aligned in palisading rows leaving anuclear zones termed Verocay bodies (Fig. 6b). The tumor was diagnosed as a benign schwannoma. Special stains for S100 (immunohistochemistry) could not be done because of unavailability of these stains in our institute. Postoperatively, the patient did well. The sutures were removed after a week, and the patient was on regular follow-up. There have been no recurrences.

DISCUSSION

Neurilemmomas or schwannomas are rare ectodermal tumors of the peripheral nerve sheath that arises from Schwann cells [1]. A schwannoma is typically solitary, well-circumscribed, encapsulated, slow-growing tumor that develops from a single fascicle within the main nerve and abuts the rest of the nerve, eventually causing neurodeficit. They are mostly benign in nature and rarely cause symptoms. They are commonly seen in the eighth cranial nerve, where they are associated with neurofibromatosis Type 2, knowing to cause compressive symptoms. They are mostly asymptomatic and may be detected on incidental imaging. In certain cases, they may be symptomatic due to pressure over the surrounding nerves.

Imaging has a major role identification of such nerve sheath tumors. Schwannoma may have a fusiform shape, located eccentrically in relation to the nerve, compared to neurofibroma, which is central and inseparable from its parent nerve. Schwannoma shows intense enhancement with contrast, while the "ancient" schwannoma would enhance non-uniformly but shows marked hyperintensity on T2 sequence characteristic

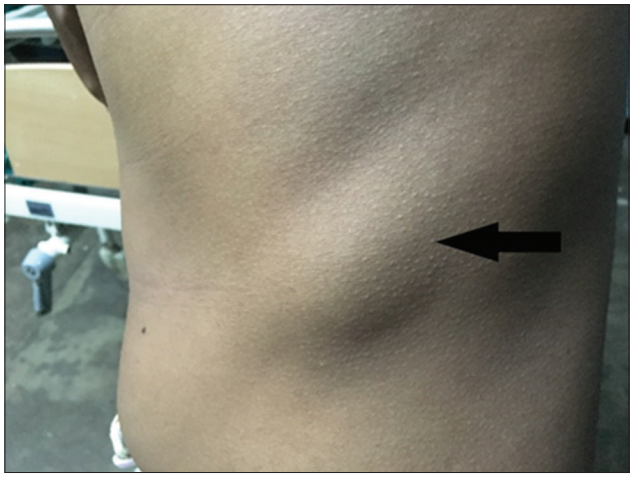

Figure 1: Lateral view of the patient showing the mass in the left flank (black arrow)

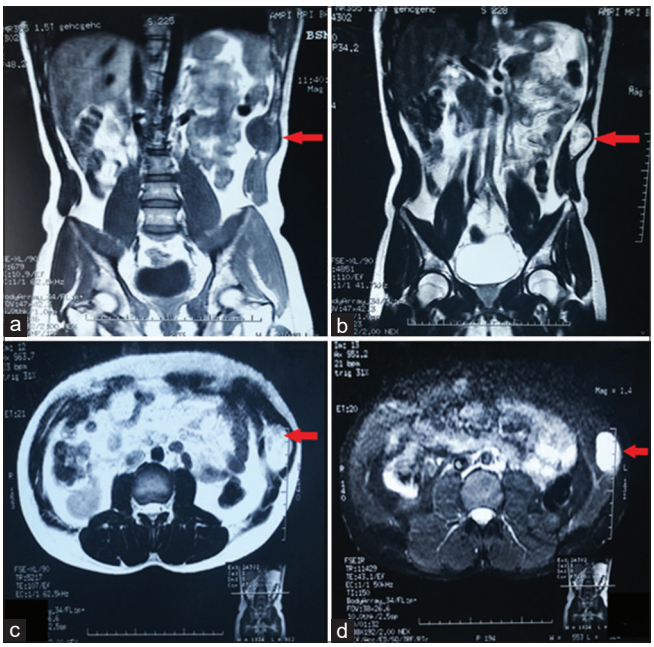

Figure 3: T1-weighted (T1WI) coronal magnetic resonance imaging (MRI) showing (a) a iso to hypointense lesion (red arrow). (b) A heterogeneously hyperintense lesion (red arrow) on T2WI axial MRI. (c) A heterogeneously hyperintense lesion (red arrow) on T2WI coronal MRI. (d) MRI with fat suppression (inversion recovery) sequence axial view showing hyperintense mass over the abdominal wall (red arrow)

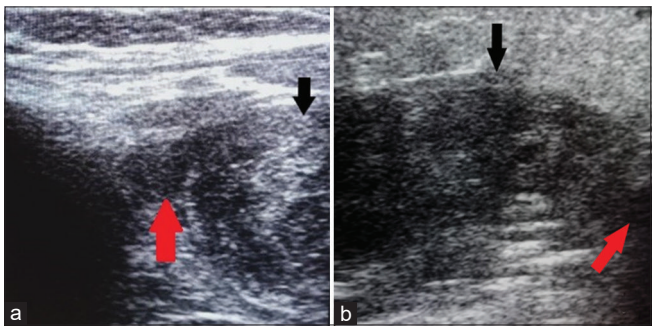

Figure 2: (a and b) Ultrasound well-defined, encapsulated (black arrow), ovoid heterogeneously hypoechoic space-occupying lesion within the muscle plane, with a hypoechoic extension (red arrow)

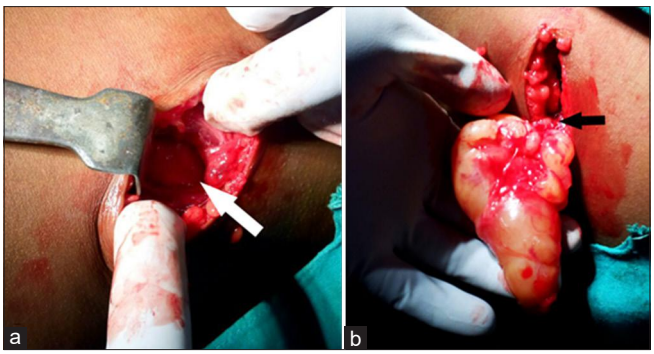

Figure 4: (a) Intraoperative view of the tumor (white arrow). (b) Resected tumor - nerve entering the tumor (black arrow)

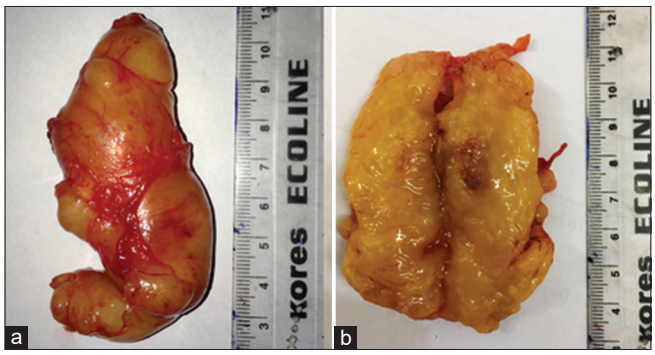

Figure 5: (a) Gross view of the resected specimen. (b) A longitudinal section of the specimen showing the pale yellow color of the tumor

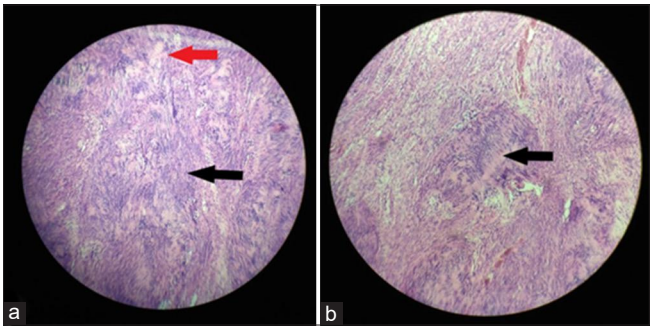

Figure 6: Histopathological image showing (a) hypercellular areas (Antoni A) (black arrow) with Verocay bodies and a hypocellular area (Antoni B) (red arrow) and (b) palisaded nuclei with anuclear zones (Verocay bodies) (black arrow)

of cystic degeneration patient. A few characteristic signs may be seen on MRI as described by Kakkar *et al*. The "split-fat sign" refers to the presence of fat on the upper and lower poles of a lesion on T1WI, which suggests the intermuscular location of the lesion. The "fascicular sign" is represented by multiple ring-like structures, which appear as hypointense foci within the hyperintense area on T2WI, possibly reflecting the fascicular bundles seen histologically [5]. Histopathological features include an encapsulated tumor with spindle cells with focal nuclear palisading arranged in dense (Antoni A) and loose (Antoni B) areas. Connective tissue fragments (Verocay bodies) and intranuclear vacuoles are often present. The "ancient" adjective is added to a Schwannoma when it contains degenerative changes such as cysts, calcification, interstitial fibrosis, vascular hyaline degeneration, and hemosiderin deposition. A suspicion of malignancy may arise when there are nuclear atypia and hyperchromatic cells. Schwannomas react positively with S100 on immunohistochemistry, and this also helps to distinguish it from malignancy. The treatment is complete excision of the tumor to avoid recurrences [6].

Various authors have reported their experiences with the abdominal wall schwannomas. In 2016, Ginesu *et al*. [7] from Italy reported a case of a 62-year-old female referred with a right iliac fossa pain that revealed a well-circumscribed mass, with small calcifications. In 2015, Balzarotti *et al*. [8] reported a case of symptomatic schwannoma of the abdominal wall from Switzerland. In 2013, Mishra *et al.* [9] reported another similar case from Libya. In 2010, Bhatia *et al*. [10] reported an incidental benign ancient schwannoma after a privately funded computed tomography scan in a 64-year-old woman who had to undergo surgery for the tumor, from Essex.

CONCLUSION

There is a dearth of literature that highlights the behavior and nature of ancient schwannomas occurring in the abdominal wall. To conclude, we state that most of these tumors may be asymptomatic. A multidisciplinary approach must be taken to approach these tumors. Radiological imaging helps the surgeon to diagnose the tumor and a complete resection is essential to prevent a recurrence.

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REFERENCES

1. Hajdu S. Peripheral nerve sheath tumors histiogenesis, classification, and prognosis. Cancer 1993;72:3549-52.
2. Maneschg C, Rogatsch H, Bartsch G, Stenzl A. Treatment of giant ancient pelvic schwannoma. Tech Urol 2001;7:296-8.
3. El-Barhawy M, Khoubehi B, Hrouda D. Perirenal schwannoma: A case report. J Med Case Rep 2008;2:189-94.
4. Ninos A, Douridas G, Liapi G. Schwannoma in the inguinal canal masquerading an inguinal hernia. Hernia 2004;8:73-5.
5. Kakkar C, Shetty CM, Koteshwara P, Bajpai S. Telltale signs of peripheral neurogenic tumors on magnetic resonance imaging. Indian J Radiol Imaging 2015;25:453-8.
6. Kumar V, Abbas AK, Aster JC. Robbins and Cotran Pathologic Basis of Disease. 9th ed. Philadelphia, PA: Elsevier/Saunders; 2015. p. 1246-9.
7. Ginesu GC, Puledda M, Feo CF, Cossu ML, Fancellu A, Addis F, *et al*. Abdominal wall schwannoma. J Gastrointest Surg 2016;20:1781-3.
8. Balzarotti R, Rondelli F, Barizzi J, Cartolari R. Symptomatic schwannoma of the abdominal wall: A case report and review of the literature. Oncol Lett 2015;9:1095-8.
9. Mishra A, Hamadto M, Azzabi M, Elfagieh M. Abdominal wall schwannoma: Case report and review of the literature. Case Rep Radiol 2013;2013:ID 456863.
10. Bhatia RK, Banerjea A, Ram M, Lovett BE. Benign ancient schwannoma of the abdominal wall: An unwanted birthday present. BMC Surg 2010;10:1-5.

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