Retroperitoneal schwannoma and challenges for pre-operative diagnosis: A case report

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SUMMARY

Schwannomas are rare neoplasms with non-specific clinical features which may render diagnosis difficult. Ideally, complete surgical excision is preferred, as malignant schwannoma lacks sensitivity to chemo and radiotherapy. We report a 40-year-old female who was referred to our tertiary referral hospital for ultrasound-guided biopsy of a non-palpable left pelvic mass with normal tumour markers and previous history of diffuse large B-cell lymphoma who completed 10 cycles of chemotherapy, with follow-up revealed no bone marrow infiltration by the primary disease. Serial computed tomography suggested retroperitoneal mass while her magnetic resonance imaging of the pelvis revealed a well-encapsulated multiloculated solid cystic mass. The definitive diagnosis was only confirmed by post-operative histopathological report.

INTRODUCTION

Schwannoma tumours originate from the sheath of central or peripheral neural cells. Retroperitoneal schwannoma is a rare tumour, and it is usually detected incidentally. It develops between the second and fifth decades of life with a slightly higher morbidity in women than in men.[1]

Schwannomas comprise 0.3–3.0% of retroperitoneal tumours. Due to the large and accommodating retroperitoneal space, the diagnosis is often delayed, resulting in a significant large-sized lesion with compression symptoms and displacement of adjacent structures. Schwannomas can be found in the head, neck, and flexor surfaces of the extremities too. Detection of a malignant schwannoma is important, as it will affect the treatment strategy. Some surgeons advocate attaining negative margins with en bloc excision as malignancy cannot be ruled out intraoperatively even if frozen section is used.[2] We report a case of retroperitoneal Schwannoma which was visualised as pelvic mass by ultrasound, CT and MRI; however, confirmed diagnosis was only after resection based on histopathological examination and immunohistochemistry staining.

CASE REPORT

A 40-year-old woman parity 4 presented with acute abdomen in July 2019. Abdominal ultrasound revealed intra-abdominal fluid collection. Exploratory laparotomy revealed multiple small bowel mesenteric lymph nodes and drainage of 400 ml intraperitoneal fluid. Nodule excision biopsy confirmed diffuse large B-cell lymphoma (DLBCL) grade 3A and intraperitoneal fluid cytology reported no malignant cells.

CT is a good imaging modality to assess retroperitoneal masses in view of providing sectional images of the mass and retroperitoneal structures.[3]

Subsequent CT scan showed left lateral pelvic hypodense lesion (5.7 x 7.0 cm) with multiple mesenteric nodes (Figure 1a). She had a total of 10 cycles chemotherapy (CHOP, R-CHOP, R-ESHAP). Bone marrow aspiration trephine showed no bone marrow infiltration thereafter. In November 2020, she developed peripheral neuropathy which was attributed to chemotherapy. Repeated CT in December 2020 showed all lymphadenopathies were stable while left pelvic wall heterogenous enhancing lesion (7.6 x 6.0 cm) was marginally increased in size (Figure 1b).

In January 2021, patient was referred for ultrasound-guided biopsy which revealed fibrofatty tissue, not representative of the mass. Bimanual examination revealed a smooth non-ballotable tender mass lateral to the left adnexa. Ultrasound showed (7.0 x 8.0 cm) multiseptated left adnexal mass with solid area and no ascites. Her tumour markers cancer antigen 125, carcinoembryonic antigen, alpha-fetoprotein, cancer antigen 19-9, beta human chorionic gonadotropin, and lactate dehydrogenase were all normal.

MRI may discriminate retroperitoneal schwannoma from other retroperitoneal tumours when compared with ultrasound and CT.[4]

Pelvic MRI suggested an ovarian mass - a well-encapsulated multiloculated solid cystic mass (predominantly cystic), with internal septations. Heterogeneously hypointense with T1W sequence (Figure 2a) and hyperintense with T2W (Figure 2b). In view of such ovarian mass and the possibility of cancer recurrence or transcoelomic metastasis, we had decided for exploratory laparotomy and found a large left obturator mass (8.0 x 6.0 cm) extending deep into the pelvis and sacral plexus. Since the tumour was adherent to the iliac vessels and

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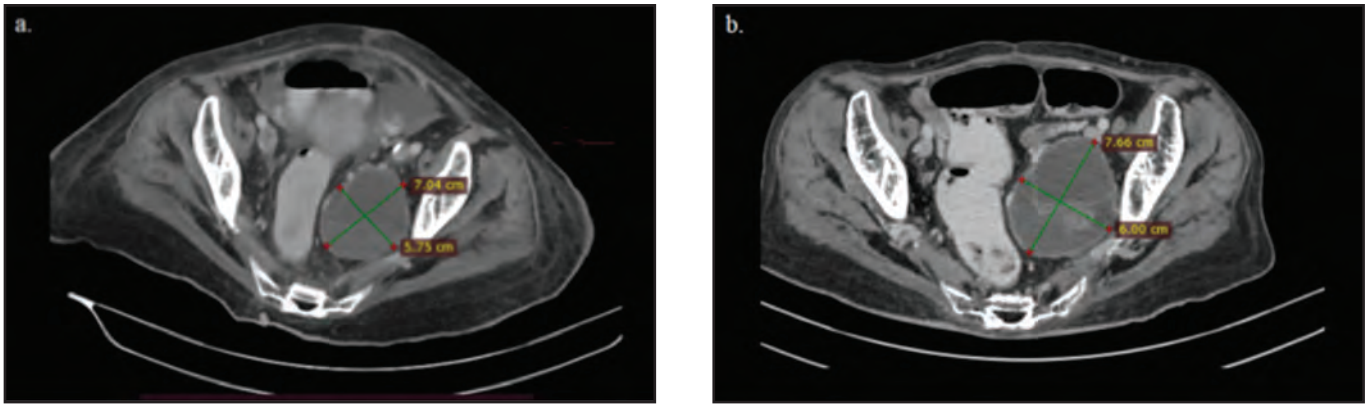

Fig. 1: CT showing (a) left lateral pelvic hypodense lesion (5.75 x 7.04 cm). (b) multilobulated heterogenous left pelvic enhancing lesion (7.66 x 6.00 cm).

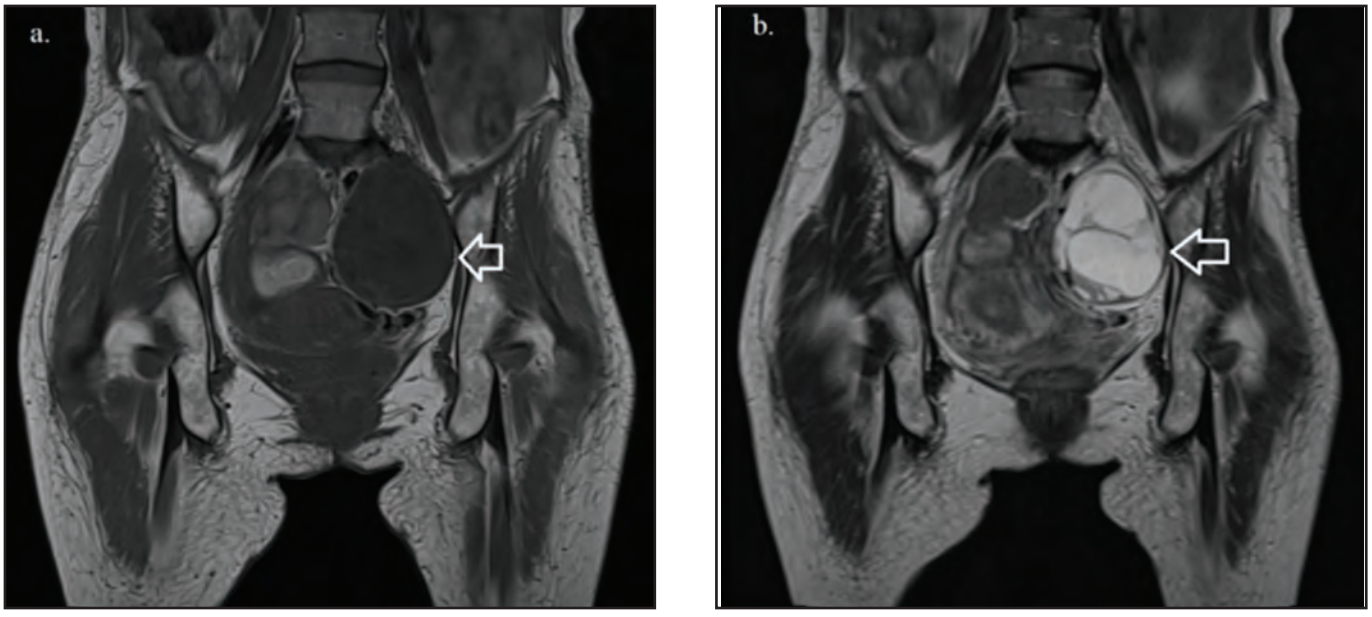

Fig. 2: MRI shows well-encapsulated predominantly cystic mass. (a) Heterogeneously hypointense lesion with T1W sequence (arrowed). (b) Heterogeneously hyperintense lesion with T2W sequence (arrowed).

nerve, we had decided for R2 resection, leaving about 20% tumour remnant. Both ovaries and uterus were normal intraoperatively. Histopathological report came back as low to intermediate grade spindle cell lesion favouring schwannoma. No lymphoid tissue is present. Immunohistochemistry staining showed diffuse positivity to S100 protein (Figure 3a), negative to CD34 (cluster of differentiation) (Figure 3b), smooth muscle antibody (Figure 3c), and desmin (Figure 3d).

Our patient was discharged postoperatively well, further three monthly clinical follow-up with serial ultrasound scanning showed no growing pelvic masses.

DISCUSSION

The clinical features of benign retroperitoneal schwannoma are non-specific; with possible displacement of retroperitoneal structures. Palpable abdominal mass and abdominal pain tend to be the most common presenting complaints in advanced stage.[4] Atypical recurrent renal colic, haematuria, headache, and secondary hypertension are reported.[4]

CT is a good imaging modality to assess retroperitoneal masses in view of providing sectional images of the mass and other retroperitoneal structures,[3] while MRI may discriminate retroperitoneal schwannoma from other retroperitoneal tumours when compared with ultrasound and CT.[4] From the MRI, target sign of schwannoma can be seen as T2W1 hyperintense rim surrounding a central low signal area and fascicular sign with hyperintense multiple small ring-like structures representing the bundles within the nerves.[5] Schwannoma may present as a completely cystic lesion due to secondary degenerative changes, mimicking an ovarian cystic mass.[6] In our patient, CT scan showed multilobulated heterogenous left pelvic mass (Figure 1a and 1b) and MRI showed well encapsulated predominantly cystic mass which

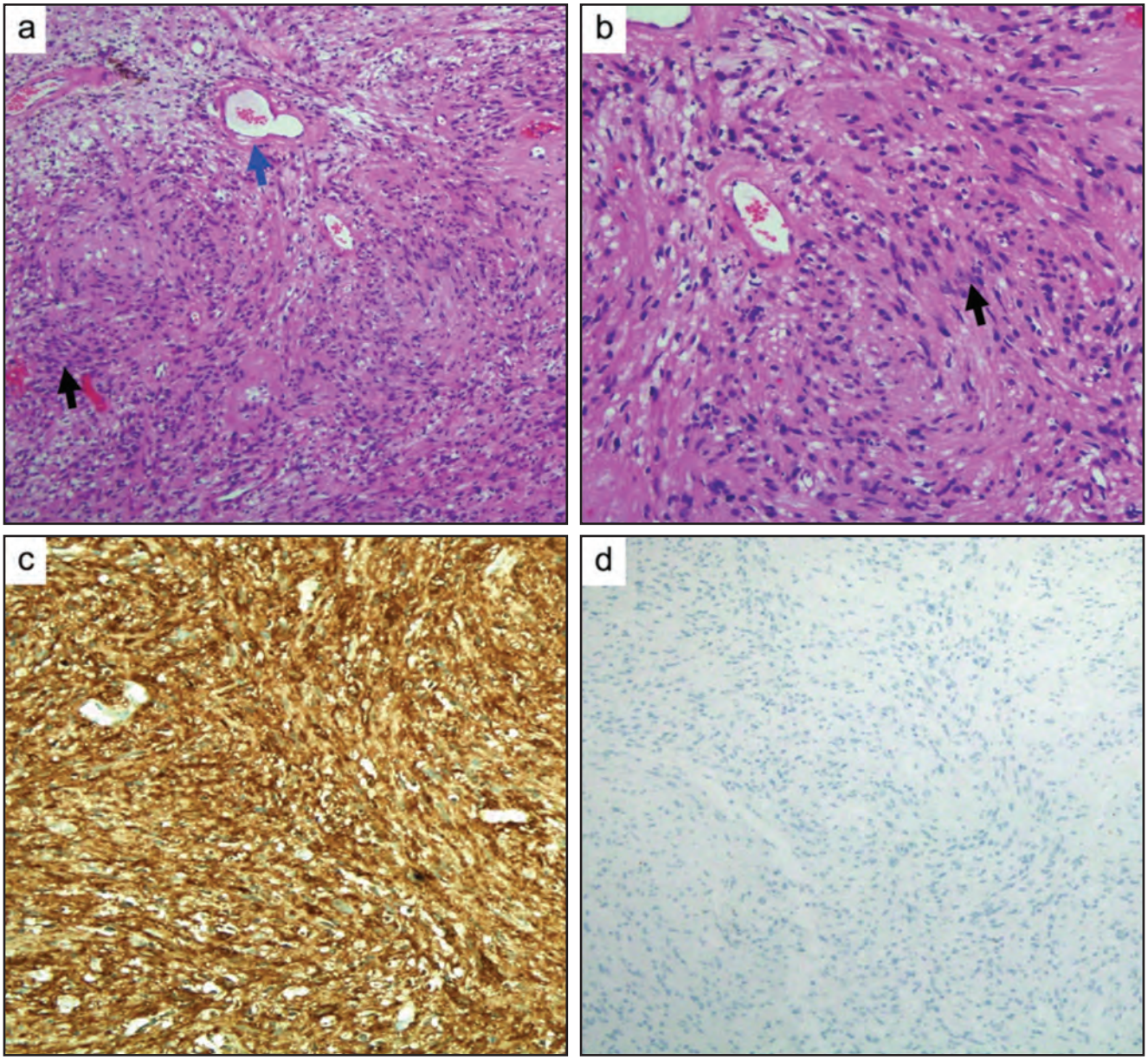

Fig. 3: (a) Microscopic examination of the lesion, biphasic characteristic of compact hypercellular, Antoni A (black arrow) and myxoid sparsely cellular, Antoni B (arrowhead) with thickened hyalinized vessels (blue arrow) within the lesion (H&E stain, 40x magnification). (b) The cells display spindle to ovoid, bland nuclei, inconspicuous nucleoli and ill-defined cytoplasmic border. Nuclear palisading around fibrillary process (Verocay bodies) is seen (arrow) (H&E stain, 200x magnification). (c) The neoplastic cells are positive for S100 (S100 stain, 100x magnification). (d) The neoplastic cells are negative for CD34 (CD34 stain, 100x magnification).

showed hypointense on T1W sequence (Figure 2a) and hyperintense on T2W (Figure 2b) with imaging suggestive of haemorrhage within the mass. In view of our patient with the previous history of DLBCL, additional Positron Emission Tomography alongside CT scan might help for further assessment of the mass.

Retroperitoneal schwannoma with cystic degeneration is histologically characterized by verocay body formation with alternating Antoni A and Antoni B areas.[6] The histopathological evaluation for our patient's mass showed spindle cell lesion suggestive of schwannoma in view of the presence of characteristic biphasic lesion of compact hypercellular Antoni A and sparsely cellular, Antoni B (Figure 3a,b) as well as strong positivity for S100 stain (Figure 3c). In our case, immunohistochemistry stained negative to CD34 (Figure 3d) showing further support to the diagnosis. The histological diagnosis of malignant peripheral nerve sheath tumours lacks standard diagnostic criteria and grading. A total of 56% of malignant peripheral nerve sheath tumours showed complete loss of staining, while 17% showed mosaic staining and 28% showed intact staining.[7] It is important to exclude malignant retroperitoneal schwannoma preoperatively for optimal treatment and

favourable outcome. Immunohistochemistry positive staining for S100, and negativity for CD34 are reliable markers for diagnosis.[8]

Complete surgical excision is the preferred curative management as the recurrence rate after enucleation or partial excision range between 16% and 54%.[9] Due to the insensitivity of malignant schwannomas to chemotherapy and radiotherapy, adjuvant therapy is not recommended and surgical re-resection is recommended for recurrent cases and longer follow-up is needed to assess the true recurrence of these tumours.[10] Our reported patient will need life-long close follow-up as R2 resection had been performed.

CONCLUSION

Diagnosis of schwannoma tumour when it presents in the retroperitoneal area needs high suspicion and good clinical judgement. As retroperitoneal schwannoma is rare, it should remain as a differential diagnosis for any retroperitoneal mass. Patients with previous history of malignancy need to be investigated extensively especially if the tumour is located intra-abdominally. The location of a retroperitoneal lesions poses high surgical risk; therefore, resection should be based on radio-surgical consultation. Histological confirmation is the definitive diagnostic method. Clinicians should be aware that retroperitoneal schwannoma may mimic cystic ovarian carcinoma even in young women. It is easy to misdiagnose a pelvic mass as an ovarian tumour when it is in fact a tumour from another origin.

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DECLARATION OF PATIENT CONSENT

The authors certify that they obtained a consent from the patient to publish the images and clinical information without mentioning her name or initials.

CONFLICT OF INTEREST

There is no conflict of interest.

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