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Incidental Detection of Retroperitoneal Schwannoma: A Case Report

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Abstract

Schwannoma is a benign tumor arising from peripheral nerve sheath cells. It is rare to find them outside head, neck and extremities. We report a rare case of an incidentally detected large retroperitoneal mass in a 62-year-old male by radiological investigation, who underwent fine needle aspiration cytology which was insignificant and proceed for excision, which was found to be schwannoma on histopathological examination.

Keywords: Retroperitoneal schwannoma; Peripheral nerve; Malignant; Cancer

Introduction

Schwannomas represent an uncommon subtype of soft tissue tumors, originating from peripheral nerve sheaths. 90% of schwannomas are seen in the head and neck and flexor aspect of upper and lower limbs. Retroperitoneal schwannomas are rare, accounting for less than 10% of cases [1,2]. Incidence of benign schwannomas is 3-3.2%; in contrast, malignant subtypes occur in retroperitoneum is 1.7% of the cases [3-5]. Retroperitoneal schwannomas most of times are larger and have a higher tendency to be complicated by spontaneous degeneration and hemorrhage [6]. In this present article, we describe an incidentally detected retroperitoneal mass on ultrasonography of abdomen in 62 year old male patient.

Case Report

A 62-year-old gentleman with no previous major health problems was referred to our department with the findings of an incidentally detected retroperitoneal mass. There was no history of pain abdomen, awareness of lump, anorexia, weight loss and fever. The physical examination revealed an evident mass in the right upper abdomen. He denied any history of fever, anorexia, and asthenia or weight loss. His past medical and surgical history was unremarkable.

Per abdomen examination revealed a hard, smooth, immobile, non-tender lobular mass around 10 × 8 cm in size in left lumbar region. General physical examination was unremarkable for café-au lait spots or other features of Recklinghausen`s disease. On laboratory tests, the liver function tests, renal function test and haemogram were normal. As following Cancer Antigen 19-9 was 5 µ/ml (reference range: below 35 µ/ml.) and Cancer Embryonic Antigen (CEA 1.0 ng/ml (reference range: below 2.5 ng/ml in Non-smokers). On radiological investigations, ultrasonography of abdomen revealed a rounded lobulated heterogeneous mass with cystic areas and calcification on the left in the para-aortic region below the level of left kidney measuring 110 × 83.1 mm in size.

On further radiological evaluation, dynamic contrast enhanced computed tomography showed a well-defined rounded heterogeneous mass with predominantly solid component and soft tissue attenuation in the retroperitoneum, located in the left para-aortic region between the aorta and left psoas muscle measures 107 × 90 × 90 mm, there was no loss of fat planes between the paravertebral space and the mass. Small cystic areas and foci of calcification were seen within it. Post contrast scan reveal enhancement of solid areas (Figure 1).

Fine needle aspiration cytology of the lesion was done to know the nature of cells for differentiation of malignant from benign cells as a routine protocol followed in our institute, which revealed mature adipose tissue fragments, few muscle fragments and scant fibrous stroma in a haemorrhagic background.

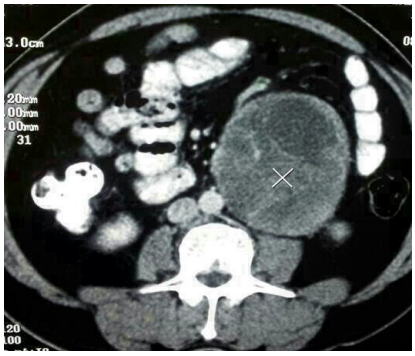

Figure 1: Contrast enhanced computer tomography abdomen showed a 107 × 90 × 90 mm well-defined rounded heterogeneous mass with predominantly away from solid soft tissue attenuation values in the left para-aortic region (white cross).

Intraoperatively, a tumor measuring 10 × 9 × 5 cm, adherent to left ureter and lumbar spine was found. There were no liver metastasis/omental/peritoneal/pelvic deposits. Complete tumor excision was done from the surrounding structures preserving the vascular structures and the left renal vein. The mass neither encased the superior/ inferior mesenteric artery or vein, nor the left renal vein. The patient`s postoperative course was uneventful and was discharged on post-operative day [7]. Histopathological analysis demonstrated Spindle cell shaped tumor cells with mild nuclear pleomorphisim, well formed Verocay bodies, hyalinized blood vessels, cystic changes and few foci of micro-calcification and peripheral lymphoid aggregates and giant cells (Figure 2). Immunohistochemical stains were positive for S-100 (Figure 3). Overall feature were suggestive of schwannoma.

Discussion

Schwannomas are benign tumors that develop from Schwann cells of the peripheral nerve sheath derived from the neuroectoderm. They

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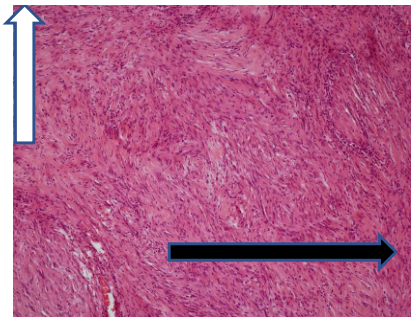

Figure 2: Shows Antoni type A (white arrow) and B cells (black arrow).

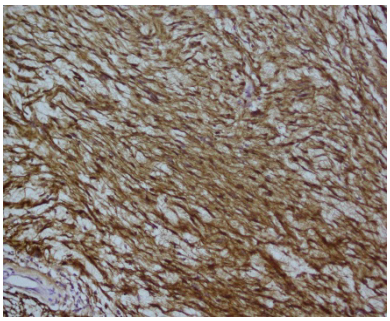

Figure 3: Shows Immunohistochemical stains were positive for S-100.

are most commonly located in the head and neck region (44.8%), upper limbs (19.1%), and lower limbs (13.5%). Retroperitoneal schwannomas are rare, accounting for only 1.2% of all retroperitoneal tumors. These tumors are frequently seen in adult population between the ages of 20 and 50. Women are affected twice as often as men. Retroperitoneal schwannomas are most often detected incidentally and may present with vague symptoms. Abdominal ultrasonography is a simple and non-invasive method for early detection of these tumors. On CECT abdomen, retroperitoneal schwannomas are visualized as well circumscribed rounded mass with heterogeneous contrast enhancement due to cystic and haemorrhagic changes and calcification. Cystic changes commonly occur in big tumours and 40% of schwannomas have cystic components. MRI is regarded as the diagnostic modality of choice for evaluation of retroperitoneal tumors due to its better delineation of the origin, extent, and internal composition of these lesions. Schwannomas are seen as hypointense on T1 and hyperintense on T2 weighted MR images. A few well-known typical imaging characteristics for schwannomas which are target sign and fascicular sign are not frequently seen in retroperitoneal schwannomas. The "fascicular sign" is appearance of bundles which is a general property of neurogenic tumors and the "target sign" is the presence of hypointense centre and hyperintense periphery on T2 weighted MRI [8,9]. CT-guided core biopsy and fine needle aspiration have been founded to be unreliable for the diagnosis of retroperitoneal schwannoma. There may be a risk of haemorrhage, infection, and tumor seeding, thus many authors do not recommend CT-guided biopsy [10]. Hence, there is no gold standard pre-operative diagnostic modality for retroperitoneal schwannoma [3-5].

A definitive diagnosis is based only on pathological, histological, and immunohistochemical findings. Schwannomas histologically consist of compact cellular lesions (Antoni type A tissue) and loose, hypocellular myxoid lesions with microcystic spaces (Antoni type B tissue). In addition, almost all schwannomas show intense immunohistochemical staining for S-100 protein, confirming the neuroectodermal origin of the tumor cells. The malignancy rate of schwannomas range from 1.7 to 30.7%, and the relapse rate is as high as 61%. Even in pathologically benign cases, relapse rate is 4.3%, and the malignant transformation rate is 12% [6-9].

The only definitive treatment for schwannoma is surgical excision as they are chemo-radiotherapy resistant. However, the necessity for negative soft tissue margins is controversial especially when adjacent tissue or viscera need to be sacrificed. The prognosis of benign schwannomas is good and the most frequent complication is recurrence of the tumor, probably due to incomplete excision, which accounts for 5–10% of cases [11,12]. Our patient is free of symptoms, recurrence or malignant transformation of tumor after one and half year of follow-up which was done by clinical examination and contrast enhanced ultrasonography [10-12].

Conclusion

We present a rare retroperitoneal schwannoma in male which was detected incidentally. Most of them are benign and requires a high index of suspicion. In this case study we found that radiological investigations plays a major role in management of patient and during follow up period. Diagnosis is confirmed by histopathology and total excision of benign tumor has a good prognosis and requires a regular follow up to rule out recurrence and as well malignant transformation.

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