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Retroperitoneal schwannoma: a case report review and literature

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ABSTRACT: Retroperitoneal schwannomas are rare, usually benign tumors that originate in the neural sheath and account for only a small percentage of retroperitoneal tumors. We report a case of a 34-year-old male presented to our department complaining of left abdominal mass and pain. An abdominal computed tomography (CT) scan showed a large retroperitoneal mass of 15 cm in diameter**.** Histopathological examination confirmed the diagnosis of benign schwannoma. After two years, at follow-up the patient was free of disease. Retroperitoneal schwannoma is a difficult clinical diagnosis and requires a high index of suspicion. Magnetic resonance imaging scan may help in the diagnosis of schwannoma and diagnosis is based on histopathological examination and immunohistochemistry. Complete excision has a therapeutic effect and a good prognosis.

KEYWORDS: Schwannoma, retroperitoneal, complete excision.

INTRODUCTION

Neural sheath tumors are a subclass of soft tissue neoplasms that include both benign and malignant schwannomas and neurofibromas. Schwannomas are found most commonly in cranial and peripheral nerves and occur rarely in the retroperitoneum, the last comprising about 3% of all schwannomas [1,2,3]. Schwannomas constitute approximately 4% of all retroperitoneal tumors. They are typically solitary, circumscribed and encapsulated lesions on gross appearance. Histologically, schwannomas are distinguished by the presence of areas of high and low cellularity, called Antoni A and B tissue, respectively [1]. They are slow growing and may produce vague local symptoms, but are usually diagnosed incidentally. They are an uncommon cause of a retroperitoneal mass, and are classically encapsulated, highly vascular and have a distinctive radiological appearance [4].Retroperitoneal schwannomas are usually larger and have a higher tendency to undergo spontaneous degeneration and hemorrhage [5]. Herein, we present a case of a 34-year-old male with retroperitoneal tumor diagnosed as schwannoma.

CASE PRESENTATION

A 34-year-old male with no previous major health problems, presented to our department complaining of left abdominal lump & pain. The condition had started nine months before. The physical examination revealed an evident mass in the left flank abdomen. He denied any fever, anorexia, asthenia or weight loss. This pain was non-radiating and did not respond to over the counter analgesics. Normal bowel habits, His initial laboratory reports were within normal limits. Computed tomography (CT) scan revealed a 13.5x12x11cm heterogeneous well limited retroperitoneal cystic mass located on the left psoas muscle and closely attached to the posterior wall of the vertebra **(Fig.1).**CT angiography revealed a 113x140x117mm, heterogeneous, necrosed mass located on the left psoas muscle and vascularized by tributaries collateral branches of abdominal aorta. This mass repulses at the top the left kidney and arrives inside at the contact of the aorta **(Fig.2)**.

An exploratory laparotomy was performed using a midline incision. The large retroperitoneal solid mass occupying left flank was found and attached to the posterior wall of the lumbar vertebra **(Fig.3a).** The tumor was encapsulated and hyper vascularized with carefully dissection, complete resection of the tumor was performed **(Fig.3b).** Histopathology confirmed

the diagnosis of an ancient schwannoma.Rare mitoses were seen, however, there were no overt features of malignancy. All margins were negative and postoperative recovery was unremarkable and patient discharged on postoperative day 7. The patient was free recurrence at 18 months follow-up.

DISCUSSION

Schwannomas (neurilemmoma) are nerve sheath tumors that usually affect the head, neck and the flexor surfaces of the extremities. It is rare to find a schwannoma in the retroperitoneal cavity and accounts for 0.3–3.2% of schwannomas [1,2, 3, 7].They are mostly benign in nature, more commonly occurs in adult females between 20–50 years age with a male to female ratio of 2:3[6]. Schwannomas have true capsules composed of epineurium. The tumor mass is characteristically eccentric with respect to the affected nerve. Retroperitoneal schwannomas show cystic degeneration in up to 60% of cases while calcification is seen in 23% of cases only. These changes make diagnosis even more difficult [7].The symptomatology is nonspecific and depends on the location and size of the lesion, cause of the slowly growth and the anatomical position of the tumor and my produces vague symptoms like abdominal pain and distention , that is why the diagnosis of retroperitoneal schwannomas is often delayed[3]. Preoperative diagnosis based on clinical examination is very difficult and so the role of imaging is important. Ultrasonography is a cheap modality for revealing a mass with semisolid or cystic areas, but it is not used widely due to specificity limitations [1]. Computed tomography (CT) and magnetic resonance imaging (MRI) are widely used as imaging techniques in the evaluation of retroperitoneal soft tissue tumors. The diagnostic value of CT does however appear mitigated by its limited resolution and soft tissue definition. CT images fail to adequately reproduce stroma heterogeneities, a main characteristic for ancient Schwanomas, when compared to MRI. However, addition of intra-venous Gadolinium as a contrast medium may provide enhancement of tissue in homogeneities within the tumor [8]. Today, MRI is the investigation of choice which describes about the origin, extent, and internal structure, and infiltration of the lesion. The typical diagnostic signs in MRI are “Target sign” which is hypo –intense Centre with hyper-intense periphery, and “Fascicular sign” which is seen in presence of fascicular bundles. These findings will be lacking in retroperitoneal schwannoma which makes the diagnosis challenging. Ultrasonography is useful in guided biopsies and CT scans detects malignant changes associated like bony erosions [9]. CT-guided biopsy or FNA may be helpful only if the sample contains enough Schwann cells to visualize microscopically. CT-guided biopsy is usually unreliable because of the cellular pleomorphic in areas of degeneration; hence degenerative cells may be misinterpreted as malignancy. Therefore the high risk of hemorrhage, infection, and tumor seeding, many investigators do not suggest CT-guided biopsy for the diagnosis of retroperitoneal schwannomas and recently, a minimally invasive approach was described for excision of retroperitoneal schwannomas using an endoscopic-assisted minilaparotomy [10].

Micro-scopically, the tumor is composed of elongated cells, arranged in a palisade fashion and taking one of two patterns. In Antoni type A, the cells are arranged in an organized compact pattern. In Antoni type B, the cells are scattered loosely in an oedematous matrix. Both patterns may coexist in the same tumor and malignant changes are rare [11]. Hence, we believe that for the preoperative diagnosis of retroperitoneal schwannomas, a high index of suspicion is mandatory, especially in the absence of characteristic imaging features, as in our patient. The differential diagnosis for retroperitoneal schwannomas includes other neurogenic tumors such as paraganglioma, neuroblastoma and pheochromocytoma. The treatment of schwannomas is only surgical, since, schwannomas are radiotherapy and chemotherapy resistant. However, the necessity for negative soft tissue margins is controversial especially when adjacent tissue or viscera need to be sacrificed [3]. Recurrence of the tumour is usually within six months of surgical excision, and it varies from 16% to 54%. The completeness of excision and tumour grading are the most important in ssessing the recurrence and survival. Prognosis is usually good, but related with the site of the lesion, differentiation, invasion, and excision clearance. Radical excision is considered as the best treatment for this condition but controversies exists regarding the margin clearance. Recent advances by laparoscopic excision have also been explained [9].

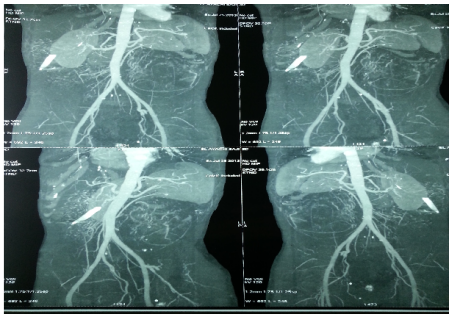


Fig. 1. CT angiography revealing a 113x140x117mm, heterogeneous necrosed mass located on the left psoas muscle and vascularized by tributaries collateral branches of abdominal aorta.

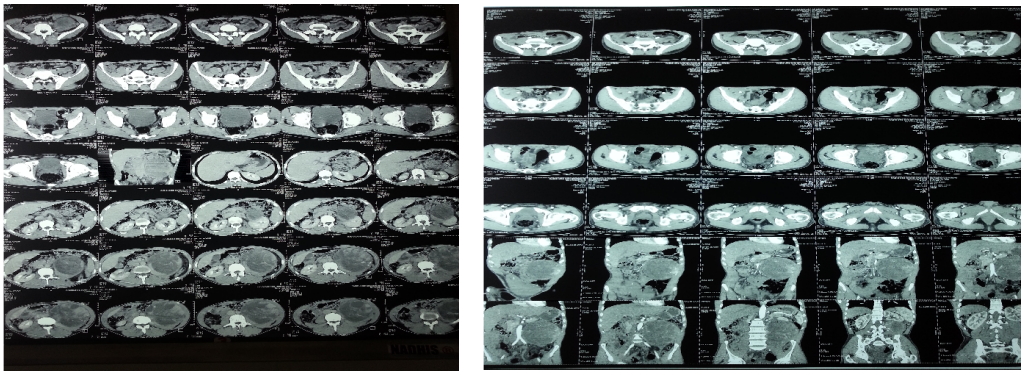
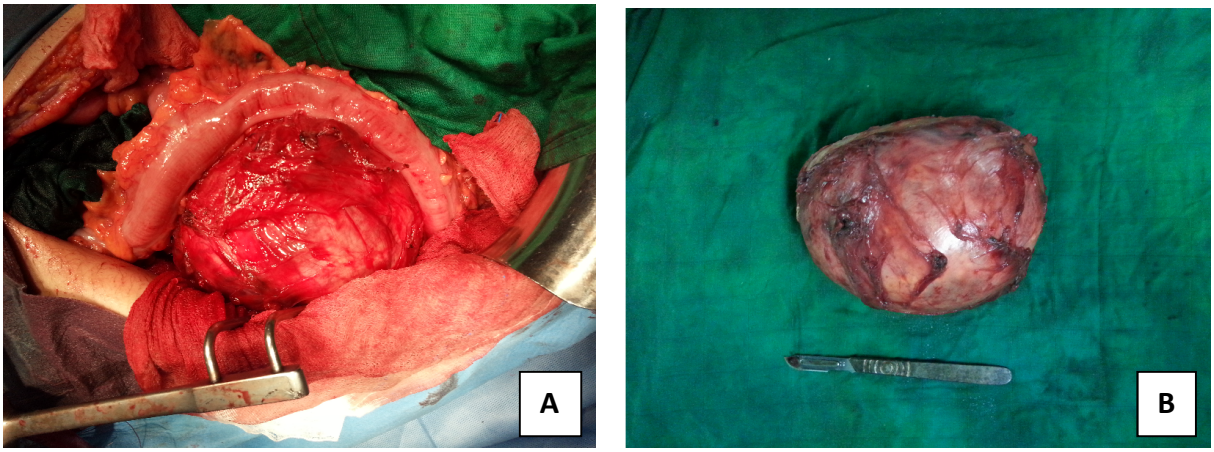


Fig. 2. CT abdominal scan shows retroperitoneal and contact with abdominal Aorta.



(Fig.3A & B).View perioperative of the tumor before and after complete resection.

CONCLUSION

Retroperitoneal schwannoma is a difficult clinical diagnosis and requires keeping in mind a high index of suspicion. Although, Magnetic resonance imaging scan may help in the diagnosis of schwannoma, the diagnosis is based on histopathology examination and immunohistochemistry. Complete excision has a therapeutic effect and a good prognosis.

CONFLICT OF INTEREST

Authors declare no conflict of interest.

AUTHOR'S CONTRIBUTION

All authors contributed to the manuscript's preparation in writing, preparing images, and literature review. All authors read and approved the final manuscript**.**

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