Retroperitoneal Schwannoma: Unusual Cause of Radiculopathy

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ABSTRACT

Schwannomas are relatively common tumors that originate from the peripheral nerve sheath. They account for 4% of all tumors found in the retroperitoneal space, with schwannomas making up approx-imately 3% of that total. The occurrence of retroperitoneal schwannoma presenting with lumbar radiculopathy is quite rare. Benign retroperitoneal schwannomas generally have a favorable outcome following complete surgical removal, with a low likelihood of recurrence, while those that are only par-tially removed have a recurrence rate of 5-10%. For a definitive diagnosis, histopathological examina-tion and immunohistochemical analysis are the most effective methods.

Keywords: Polycystic kidney disease , Retroperitoneal Schwannoma, Radiculopathy,

INTRODUCTION

Schwannoma is common peripheral nerve sheath tumors. [1,2] They are most commonly localized on the head, neck, and flexor surfaces of the extremities. Sometimes Schwannomas may appear in the posterior mediastinum but Retroperitoneal schwannoma is extremely rare, accounting for approximately 3% of all schwannomas. [2–4] Retroperitoneal schwannomas (RSs) are usually noted in the paravertebral space or presacral region.[5] Most schwannomas are benign, although malignant cases are known to occur rarely, especially if there is association with Von Recklinghausen's disease. In the absence of von Recklinghausen's disease9, these tumors rarely noted in the retroperitoneum, comprising 0.5% to 5% of all schwannomas.[2] Retroperitoneal schwannoma with polycystic kidney is not reported yet, without any other sign of von Recklinghausen's disease has rarely been reported. [9] In this report, we describe a patient of retroperitoneal schwannoma with the left lower limb radiculopathy[12] with polycystic kidney who underwent the surgical resection through retroperitoneal approach.

CASE REPORT

A 63-year-man presented with lower back pain radiating to the left lower limb for past 7 months. It was insidious in onset, progressive in nature aggravated when walking or standing and relieved when lying down. No any gastrointestinal or urinary symptoms noted. No medical history of hypertension or diabetes mellitus. No relevant familial or psychosocial history. Performing the straight leg raising test, it was found to 60 degrees on the left side while on the right side it was 90 degrees of possible elevation. The extensor hallucis longus (EHL) and flexor hallucis longus (FHL) force test showed a strength of 4/5 and 5/5 on left side respectively. Similarly, EHL and FHL on the right side was 5/5.

Sensory examination showed normal finding. Abdominal examination is unremarkable. Performed magnetic resonance imaging (MRI) of the lumbo-sacral spine revealed a relatively well-defined, smooth outlined, heterogeneously enhancing mass lesion noted along adherent to the left psoas muscle at L3 - L4 vertebral level, along the course of the left L2 / L3 nerve roots measuring ~ 2.5 x 2.9 x 3.7 cm. It shows T2 hyperintense with some central hypointense signal and T1 isointense signal change. Multiple bilateral renal cortical cysts are noted, the largest one measure ~ 6.8 x 4.8 cm in the left kidney and ~4.4 x 3.8 cm in the right kidney. (Figure 1 A, B and C). Initially, the patient was conservatively managed with oral steroids and analgesics. However, the surgical intervention was required for persistent symptoms and decreased quality of life.

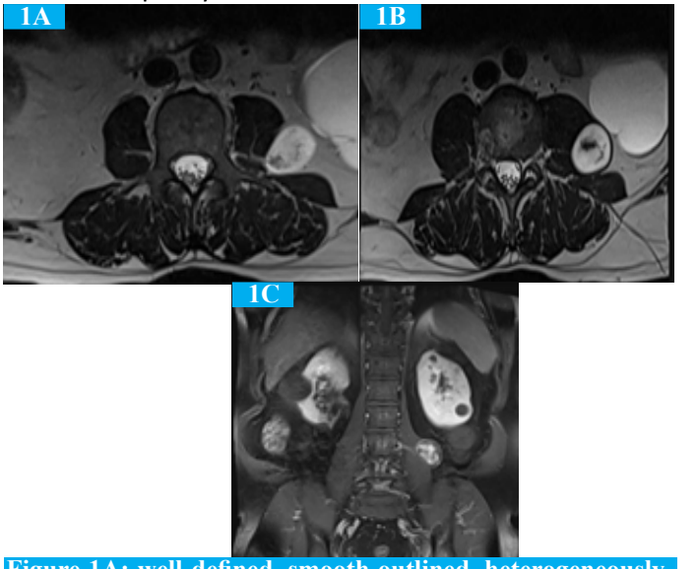

Figure 1A: well-defined, smooth outlined, heterogeneously enhancing mass lesion noted along adherent to the left psoas muscle at L3 - L4

Figure 1B: Left large renal cortical cysts with left psoas mass

Figure 1C: Multiple bilateral renal cortical cyst

The patient underwent retroperitoneal approach. Fusiform lesion identified which was arising from L4 nerve root. Lesion was separated from nerve root and gross total excision was done with preservation of involved nerve root (Figure 2A). Cyst in the lower pole of kidney identified and marsupialization also done. Histopathological report shows predominantly hypercellular and occasional hypocellular areas (Antoni A and B tissue respectively) (Figure 2B), cellular area is oval to elongated (Figure 2C) with oval to spindle shaped nuclei. Hypocellular area shows lipidized cell and lymphocytes with ectatic, hyalinized vessels and focal hemorrhage. All are compatible with Schwannoma, CNS WHO Grade 1.

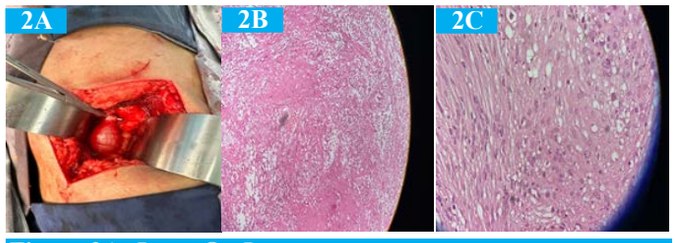

Figure 2A: Intra Op Image

Figure 2B: H & E Stain showing Hypocellular area shows lipidized cells and lymphocytes

Figure 2C: H&E stain showing cells in the cellular area are oval to elongated

DISCUSSION

Schwannomas are peripheral nerve sheaths that arise from Schwann cells and can occur in all age groups.[2,3,6,10] More than 90% of schwannomas are benign, and the most commonly located sites are the head, neck and extremities. Only 1%–3% of schwannomas are found in the retroperitoneal space. [5,7] Retroperitoneal schwannomas are mostly asymptomatic until unless they grow enough to compress surrounding structure and become symptomatic, symptoms are usually vague and non-specific. [5,11,12] Most commonly retroperitoneal mass has no limitation to grow because of large space and usually have compressive symptoms like gastrointestinal symptoms and urinary symptoms due to nearby bilateral kidney. [2,6,7] In our patient he had features of left sided lumber nerve roots compressive features and left radiculopathy.

Usually CT and MRI findings show different characteristic features such as an encapsulated, homogeneous, spherical, solitary mass on proximal nerves or spinal nerve roots, but none are specific.[8,13] They also have Secondary degenerative changes like cyst formation, calcification, hemorrhage and hyalinization.[14] MRI offer more specificity in the evaluation of the location, extension, and composition of lesions, which can help in surgical planning. However retroperitoneal mass is difficult to evaluate pre-operatively. In our case MRI shows relatively well-defined, smooth outlined heterogeneously enhancing mass lesion adherent to left psoas muscle at L3-L4 vertebral level. There are five small heterogeneously enhancing nodular region of varying size in spinal canal adjacent to nerve roots of cauda equina at L1-L5 vertebral levels, which all suggestive of Neurofibromatosis Type 1.Other diagnoses such as paraganglioma, neurofibroma, ganglioneuroma and retroperitoneal malignancies, such as malignant fibrous histiocytoma, lymphoma and liposarcoma, should be considered. [6,14]

CT-guided core biopsy and fine needle aspiration have been founded to be inconclusive. Only helpful if the sample contains enough Schwann cells to visualize microscopically. There is risk of hemorrhage, infection, and tumor seeding; thus, no recommendation for CT-guided biopsy. Therefore, surgical resection is the only appropriate approach for pathologic evaluation to diagnosis of retroperitoneal schwannoma.[15]

Surgical resection is the gold standard treatment for retroperitoneal schwannomas.[6,15] Preoperative diagnosis is challenging and a definitive diagnosis is made only by histopathological and immunohistochemical analyses of surgical specimens. As in our patient all the MRI features suggestive of Neurofibromatosis Type 1. But after the complete resection with preservation of involved nerve roots of retroperitoneum schwannoma, histological report is compatible with Schwannoma. Benign retroperitoneal schwannomas have good prognosis after complete resection with a low risk recurrence and have 5-10% of recurrence on partial resection. [6,15]

CONCLUSION

Retroperitoneal Schwannomas are uncommon benign tumors arised from peripheral nerve sheaths, and they are even less frequently associated with radiculopathy symptoms. The preferred treatment is complete surgical resection of the tumor along with decompression. The most reliable methods for definitive diagnosis are histopathological examination and immunohistochemical analysis. It is important to consider Schwannomas in the differential diagnosis of any retroperitoneal abdominal masses.

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