Retroperitoneal Intramuscular Neurilemmoma - An Unusual Presentation

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ABSTRACT

Neurilemmomas or Schwannomas originate from Schwann cells of the peripheral nerve fibers and are usually located in the head, neck, and flexor surfaces of the extremities. Primary tumors of the retroperitoneal region are quite rare and schwannomas comprise only 1–6% of them. They are insidious in onset and have nonspecific and misleading symptoms. It is difficult to diagnose them and it becomes more challenging if they undergo secondary changes. Here we present a case of 32 year old female with recurrent right abdominal pain, intermittent fever and pain in Right lower limb during walking for 6 months. Ultrasound revealed a Psoas Abscess that recurred after an initial attempt of aspiration of 30ml of clear fluid, CT diagnosis of Hydatid Cyst was made and on exploratory laparotomy a cystic swelling was found to arise from intramuscular plane of Psoas muscle which was excised along with its capsule which after Histopathological examination was found to be a Neurilemmoma. Literature review provides information about the rarity of the condition and information regarding the diagnosis and treatment of this unusual case.

Key Words: Neurilemmoma, Schwannoma, Psoas Muscle, Hydatid Cyst

Introduction

Neurilemmomas are tumors arising from Schwann cells of the peripheral nerve sheaths. They are generally slow growing and painless tumors and predominantly occur in females between the 2nd and 5th decade of life. Most schwannomas occur in the cephalocervical region and limbs, tumors in the retroperitoneal location are rare and comprise 6% of primary retroperitoneal neoplasms. The majority of retroperitoneal schwannomas are benign in nature although malignant ones have also been reported. Among all schwannomas, only 0.7% benign and 1.7% malignant ones are reported to be located in the retroperitoneal region. And among schwannomas in retroperitoneum, 98% are benign and about 2% are malignant and the incidence of malignancy is more in schwannomas of retroperitoneum than the peripheral ones. Typically, it is very difficult to diagnose retroperitoneal tumors before the operation, since both clinical and radiologic features specific to schwannomas are usually absent. Here we describe a rare case of retroperitoneal Intramuscular (Psoas Muscle) neurilemmoma.

Case Report

A 32 year old female presented with chief complaints of Recurrent RIGHT abdominal pain with intermittent fever and pain in right lower limb during walking for past 6 months. Pain started in the right lumbar region, was insidious in onset and gradually progressed in intensity with radiation to right lower limb which was exacerbated on walking and relieved on lying down with legs flexed at the knee and hip joints. She had low grade, intermittent type of fever with evening rise of temperature and without any chills and rigors and had an antalgic gait at the time of presentation without there being any previous history of trauma. No history of TB, DM and no history of similar illness in any family members.

On Examination of Abdomen

On Inspection: Abdomen was scaphoid in shape, flanks were normal, a solitary lump visible in Right Lumbar lesion approximately measuring 5x4 cm. On Palpation: A swelling of size 4x4 cm in Right lumbar region without any local rise of temperature or tenderness. With smooth surface, ill defined margins, firm consistency. It was non pulsatile, non reducible, non compressible without any impulse on coughing, didn’t move with respiration, had restricted independent mobility in both axes & didn’t fall forward on knee elbow position.

Examination of Musculoskeletal System

No Gibbus, no tenderness in spinal column, Fixed Flexion

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Deformity of Hip joint about 15 degrees.

Patient was admitted with Differential Diagnosis of Psoas Abscess and Retroperitoneal tumor, lymphoma and was subsequently investigated.

Routine hematological investigations were within normal limits, ESR & ADA were normal and the chest x-ray and x-ray spine revealed no abnormality. USG revealed findings consistent with PSOAS ABSCESS and USG guided Aspiration was done. 30ml of clear fluid was aspirated that coagulated immediately. The coagulum was sent for analysis to rule out Hydatid disease, TB & Malignancy. Microscopically the cytosmear was pauci cellular with a proteinaceous background and had no inflammatory cells or parasites. Culture yielded no growth.

Patient improved symptomatically following aspiration and was discharged and followed up at monthly interval. On 3rd monthly checkup she had recurrence of symptoms and a CT scan revealed [Figure-1] Multiloculated cystic lesion in Rt lumbar region near Rt lower pole of kidney displacing it laterally with thin & thick septations & echoes within, without any evidence of internal vascularity F/S/O Hydatid cyst. The patient was taken up for Exploratory Laparotomy under General Anesthesia and the lesion was approached through right lumbar incision. There was an INTRAMUSCULAR (Psoas Muscle) Cystic lesion of size 8x4 cm [Figure-2] which was excised in Toto and the cut sections revealed solid and cystic areas with multiple septations with areas of hemorrhage. [Figure 3]

Histopathology revealed fascicular arrangement of strand spindle cells, alternatively hypo cellular areas with marked nuclear palisading, well-formed VEROCAY body and hyper cellular areas with edema. Hallmark pattern of Antoni A & Antoni B areas were identified and a final diagnosis of INTRAMUSCULAR NEURILEMMOMA was established. [Figure-4]

Post operative recovery was normal and patient was discharged on post operative day 7 and in subsequent follow-ups there has been no evidence of recurrence.

Discussion

Schwannoma or neurilemmoma is defined as a benign neoplasm arising from the myelinated nerve sheaths. It predominantly consists of Schwann cells characterized by their palisading architecture. Schwannomas usually occur in the young and middle aged population (20-50 yrs) with a female to male preponderance of nearly 2:1. They are neuroectodermal in origin and most commonly occur in the head and neck region (44.8% of cases), upper limbs (19.1%), and lower limbs (13.5%). In contrast, it is relatively rare in the retroperitoneum, with an occurrence rate of 0.7%. Retroperitoneal region is a rare location for schwannomas except in patients having Von Recklinghausen’s disease. It is also noteworthy to mention that malignant degeneration particularly takes place in association with Von Recklinghausen’s disease. Symptomatology of benign schwannomas is highly nonspecific and depends on the location and size of the lesion. If symptomatic, the most common presenting features are vague abdominal pain, heaviness and low backache. Neurological symptoms though have been described in literature but are usually rare.

There is no gold standard diagnostic method for retroperitoneal Schwannoma; therefore, it is difficult to provide a definitive diagnosis of retroperitoneal Schwannoma before the operation. An ordinary Schwannoma is depicted as a well-defined and inhomogeneous low-density mass on CT images. On MRI, schwannomas are seen as masses with hypointensity on T1-weighted images and hyperintensity on T2-weighted images. A definitive diagnosis is based on pathological, histological, and immune histochemical findings. Histologically, schwannomas consist of compact cellular lesions (Antoni type A tissue) and loose, hypocellular myxoid lesions with microcystic spaces (Antoni type B tissue). Characteristic histological finding is the presence of VEROCAY BODY. Additionally, almost all schwannomas show intense immunohistochemical staining for S-100 protein, confirming the neuroectodermal origin of the tumor cells.

Wide surgical resection in cases of benign retroperitoneal schwannomas has been advocated by some authors based on their belief that malignancy can never be totally excluded. Others have reported that, even in difficult cases where complete removal of the tumor was impossible and simple enucleation was performed, no tumor enlargement or malignant change was observed. Wherever possible complete surgical excision must be performed for schwannomas; since, they are not sensitive to radiotherapy and chemotherapy. The prognosis of benign schwannomas is good and the most frequent complication is recurrence, probably due to incomplete excision, which is reported in only 5-10% cases.

Conclusion

Retroperitoneum is an uncommon site for neurilemmoma and INTRAMUSCULAR retroperitoneal neurilemmoma is a rarest of rare entity. Nonspecific
lower back pain might be the only presenting feature in retroperitoneal Schwannomas, causing delays in diagnosis and treatment. MRI is the investigation of choice in Preop period and Complete surgical excision of the tumor along with its capsule is the treatment of choice and excisional biopsy with HPE of the specimen is the Gold Standard for diagnosis.

**Figure - 1: CT Finding**

**Figure - 2: Intra OP Identification of the Swelling in Psoas Intramuscular Plane**

**Figure 3 : Cut Section**

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**Figure 4: Histopathology**

*Fascicular Arrangement of Spindle Cells with Palisading*

*Verrocay Body*


Oxidative stress is involved in the pathophysiological mechanisms of stroke (e.g., atherosclerosis) and brain injury after ischemic stroke. Statins, which inhibit 3-hydroxy-3-methylglutaryl coenzyme A (HMG-CoA) reductase, have both pleiotropic and low-density lipoprotein (LDL)-lowering properties.