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An Interesting Case of Ancient Schwannoma in Sciatic Nerve

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Abstract

Introduction: Schwannomas are benign, encapsulated, slow-growing and usually solitary tumors originating from Schwann cells of the peripheral nerve sheath. Schwannomas of the superficial peroneal nerves are very rare, and therefore scarcely documented in the literature. Schwannomas represents 8% of all soft tissue tumors with sciatic nerve involvement less than 1%, Sciatic nerve schwannoma is the rare cause of sciatica.

Case Report: A 50 yr. old male patient presented to surgery outpatient department with complaints of swelling in the posterior aspect of right thigh since 2 yrs., insidious in onset gradual in progression. It was associated with numbness of right leg, pain on walking, sudden increase in size in the last 3 months. Examination showed a swelling of 8 x 6 cm in posterior aspect of right thigh, firm, mobile, and not associated with peripheral vascular deficit. Imaging showed a well encapsulated heterogeneous lesion in the intermuscular plane in the posterior aspect of the right thigh, probably peripheral nerve sheath tumor. Trucut biopsy revealed features of Benign spindle cell neoplasm. Histopathology Examination revealed features of ancient schwannoma.

Keywords: Ancient Schwannoma ,Electrodiagnostic , MR Neurography , Schwann Cell , Verocay Bodies , S-100

Introduction

Schwannomas, or neurilemmomas, are benign soft-tissue tumors, arising from the Schwann cells of the peripheral nerve sheaths. They are slow growing and less likely to turn malignant. Although they are the commonest benign peripheral nerve sheath tumors, occurrence on the lower limbs account for less than 1% of all cases.

Ancient Schwannomas are rare variants of these tumors. They are termed “ancient” because of the degenerative features acquired with increasing age.

While clinically silent, they might be detected incidentally or become symptomatic due to mechanical compression and presence of pain, swelling or a lump. The diagnosis relies on clinical history, physical examination and imaging studies.

Magnetic resonance (MR) neurography offers high-resolution visualization of structural peripheral nerve tumors, hence the representing diagnostic tool of choice. The availability of MR neurography may be limited, and costs can be significant, therefore ultrasonography provides accurate imaging modality for diagnosis.

Histologically, the characteristic degenerative findings of ancient Schwannomas may be mistakenly termed malignant. This type of benign mesenchymal neoplasia usually exhibits homogeneous nuclear hyperchromasia and pleomorphism with possibly small mitotic activity.

Depending on the symptoms, surgical treatment is usually advised. Studies indicate that a neurilemmoma can be removed by delicate enucleation with an acceptable risk of injury to the nerve trunk.

Case Report

A 50 yr. old male patient presented to surgery outpatient department with complaints of swelling in posterior aspect of right thigh since 2 yrs., insidious in onset gradual in progression. It was associated with numbness of right leg, pain on walking, sudden increase in size in the last 3 months. Examination showed swelling of 8 x 6 cm in posterior aspect of right thigh, firm, mobile, and no peripheral vascular deficit and no Tinel's sign. Ultrasound Imaging showed a well encapsulated heterogeneous lesion in intermuscular plane in posterior aspect of right thigh, probably peripheral nerve sheath tumor. Trucut biopsy revealed features of Benign spindle cell neoplasm.

Imaging - MRI of right leg (Fig. 1a &b) revealed a well encapsulated heterogeneous lesion in the posterior aspect of midthigh, exhibiting intermediate signal intensity on T2 weighted images. These findings suggested that the lesion in close proximity to sciatic nerve was likely to be a peripheral nerve sheath tumor.

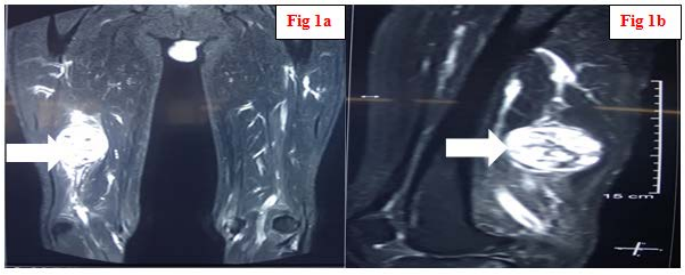

Figure 1 a and b: Magnetic Resonance Imaging showing about 6.1*5.1*4.4 cm lesion in intermuscular plane seen in coronal and sagittal plane shows continuity of mass in sciatic nerve (arrow).

Patient was taken up for surgery with a clinical diagnosis of a benign soft tissue swelling Excision under SA. intra operative findings – At surgery the well encapsulated schwannoma was enucleated from the underlying sciatic nerve. Hemostasis attained. Wound closed with drain in situ. POP applied.

Patient post-operative period was uneventful and there was no palsy. HPE showed Ancient Schwannoma.

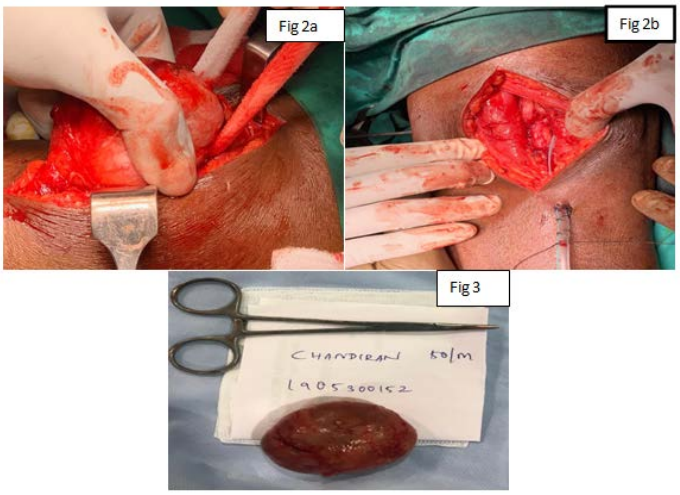

Fig 2a ,2b and 3: showing Excision of the tumor completely, Complete Enucleation of tumor with drain placed and Specimen in toto

Discussion

A Schwannoma, also known as neurilemmoma, neuroma, neurolemoma or Schwann cell tumor, is an encapsulated neoformative lesion that arises from neurilemmal cells which normally produce insulating myelin sheath covering peripheral nerves[(1)].

Schwannomas often occur in fourth and fifth decade and has a 1.6:1 female predilection. They are found in varied locations as the brachial plexus and the sciatic nerve. Rarely, Schwannomas can be found in the leg or in the foot and ankle region[(2)].

Schwannoma are slow-growing and they don't traverse the nerve but remain in sheath lying on top of it. This explains why they are clinically silent and present as an incidental finding. Patients may sometimes note cosmetic deformity or a palpable mass. An increase in size increases the compartment pressure.

The evaluation of a patient with a suspected pathologic nerve condition broadly includes history, physical examination, and imaging. Plain films may not reveal any changes whereas computed tomography (CT), magnetic resonance (MR), especially MR neurography, may display a peripheral nerve tumor in a more detailed manner. Additional diagnostic tests, including electromyography (EMG) and nerve conduction study (NCS), evaluate neuromuscular function to assess denervation, preservation of motor units, or conduction loss.

The term "ancient Schwannoma" refers to degenerative changes, which occur in these tumors with increasing duration. This rare variant was described by Ackerman and Taylor in 1951, and constitute 0.8% of all soft-tissue tumours. The histologic analysis is characterized by interstitial hyalinization, degenerative atypia, and cells with large and hyperchromatic nuclei.

The cause of these neoplasms is unknown[3]. Neurilemmoma can be associated with von Recklinghausen disease; when this is the case, multiple tumors often are present. They affect persons aged 20-50 years. Common locations for the tumors are, in order of decreasing frequency, the head and flexor surfaces of the upper and lower extremities and the trunk.

Its clinical presentation can be varied[4]. Some may involve the spinal nerve roots and present with symptoms that mimic those of herniated disk disease of the spine[5,6]. Tenderness to palpation is often present; secondary neurologic symptoms may occur if the tumor is large.

When involving the C7 nerve root, it has been described as a cause of thoracic outlet syndrome. Lesions in the sciatic nerve can mimic diskogenic low-back pain.

In extremities, neurilemmomas can present either as an asymptomatic mass (usually mobile in transverse plane and tethered along the axis of the nerve from which it arises) or as mild, localized pain and paresthesia resulting from pressure on the nerve of origin. Masses are slow-growing and can exist for months to years without producing symptoms. The average time from onset of symptoms to diagnosis is 5.5 years. Lesions in proximal nerves may cause distal symptoms. If these masses occur in well-defined compartments (eg, wrist or ankle), they can present as either carpal tunnel or tarsal tunnel syndrome.

Special studies to consider include computed tomography (CT) and magnetic resonance imaging (MRI). MRI is particularly useful ; it shows a usually round or oval mass with a moderately bright signal on T1-weighted images and a bright, heterogeneous signal on T2-weighted images. The mass is usually less than 2.5 cm in size. The lesion enhances uniformly with gadolinium contrast.

Lesions of the spinal cord often have a dumbbell shape; otherwise, they are fusiform in shape. They have an epineurium encapsulation, frequently with overlying vessels. The cut surface is pink or white. In very large masses, degenerative cysts, hemorrhage, or dystrophic calcification may be present. Neurilemmomas have a well-defined, fibrous capsule. Histologically, there are two distinct regions, as follows:

Antoni A areas - These are cellular regions with predominantly benign spindle cells in many intersecting bundles; they may palisade around eosinophilic regions that are called Verocay bodies, and they are positive for S100 staining

Antoni B areas - These are much less cellular and have a background of loose connective tissue that is myxomatous in appearance

Occasionally, a more aggressive histologic appearance may predominate, but such forms usually lack mitotic figures.

Immunohistochemistry - Acidic protein, 100% Soluble in ammonium sulfate at neutral pH. S100 protein family is multigenic group of nonubiquitous cytoplasmic EF-hand Ca2+-binding proteins, sharing significant structural similarities at both genomic and protein levels; S100 protein family has 24 known human members each coded by a separate gene; at least 19 on chromosome 1q21. This is the marker of neural tissue / lesions and melanoma. Differentiate plexiform schwannoma[7] from MPNST (weak / negative / patchy). MelanA and MART1 appear to be superior to S100 for evaluating sentinel lymph nodes for melanoma[8].

Neurilemmomas are commonly classified according to the Enneking system for benign lesions, as follows:

Grade 1 - Inactive lesions

Grade 2 - Lesions that deform the surrounding tissues but are not destructive or locally aggressive

Grade 3 - Lesions that are locally aggressive and may invade local tissues but do not have metastatic potential

Generally, neurilemmomas are grade 2 or 3.

Like most benign tumors, neurilemmomas respond well to local resection. On inspection, the nerve is usually splayed out over the lesion. The lesion is excised marginally, and the nerve fibers are spared. Interlesional resection is warranted when complete resection would result in permanent neurologic deficit. Local control is usually excellent. Effective use of sterotactic radiosurgery (SRS) for these types of lesions has been reported.

Complete surgical resection of the tumor, whilst safeguarding the surrounding structures, resulted in cure, since no damage was sustained by the underlying nerve. Most common complication of surgical treatment of neurilemmoma is initial neurapraxia; however, this neurologic deficit can be permanent, depending on the resection of neural tissue. Generally, patients tolerate resection well, with complete and rapid relief of symptoms.

If tumor is near vital nerves or blood vessels, stereotactic body radiation therapy may be used to limit damage to healthy tissue. With this technique, doctors deliver radiation precisely to a tumor without making an incision.

Schwannomas occasionally turns malignant. Malignant peripheral nerve sheath tumors (MPNST) are a group of neurogenic tumors may be sporadic or coexist with neurofibromatosis. Originates from peripheral nerves or from cells associated with the nerve sheath, such as Schwann cells or fibroblasts. They are rare neoplasm and affect 0.001 percent of the population[9]. Epithelioid subtype is rare and represents a well-recognized entity comprising of about 5 percent[10] of these tumors.

Conclusion

These benign neural sheath neoplasms are infrequent in the lower extremities according to the literature. In the differential diagnosis of nontraumatic leg pain, benign tumors, particularly Schwannomas of the peroneal nerves should be considered. Imaging studies, mainly magnetic resonance, are quite helpful in achieving a correct diagnose. Complete surgical resection by intralesional enucleation results in cure and recurrences are uncommon.

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