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A patient with leg numbness and a solitary Neurofibroma / Schwannoma at the popliteal fossa: A case report

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Abstract
Neurofibroma/Schwannoma are common peripheral nerve sheath tumors related to Schwann cell's proliferation, and are usually found in patients with neurofibromatosis type 1. Neurofibromatosis type 1, or von Recklinghausen's disease, is diagnosed when two of the following symptoms are present: Neurofibroma, flecking over the groin or axilla, café au lait spots, optic glioma and Lisch nodules [1].
There are two types of Neurofibroma that arise from Schwann cells [1]. The solitary type, which is Neurofibroma are common peripheral nerve sheath tumors. Symptoms vary according to the site, including skin discoloration, disfiguration or neurologic deterioration. Neurofibroma cannot be seen on plain X-ray film unless they involve the bone and cause intraosseous lytic lesions. Ultrasonography plays an important role in the examination of soft tissue mass because it is inexpensive and has the advantage of being real-time for initial screening. In this study, we present a patient with a palpable mass over her right popliteal fossa. The texture of the mass was solid, without pulsation or fluctuation during palpation examination. There was no bruit on auscultation. Ultrasonography showed a well-defined, oval, solid and hypoechoic mass containing anechoic areas. Magnetic resonance imaging revealed the location of the tumor enveloped by the tibial nerve. An electro diagnostic study supported the impression of entrapped neuropathy at the right popliteal area. After surgical excision of the mass, pathological findings confirmed the diagnosis of Neurofibroma.

Keywords: Neurofibroma, popliteal fossa, solitary, ultrasonography

Introduction
A 35-year-old woman presented with a solitary mass in the right popliteal fossa, and this mass had progressively developed in the preceding 2months. The skin over the lesion was intact but had mild protrusion, as the mass was fixed to deep tissue. It did not pulsate or fluctuate. There was no bruit on auscultation. This lesion was initially painless, but tenderness with radiation to the right leg and volar foot had been noted in the preceding 6 months. The patient also experienced persistent numbness over the right calf region in the later 2 months. Other physical examinations revealed a positive Tinel's sign, mild weakness of the right gastrocnemius and decreased Achilles reflex, with normal strength of the right tibialis anterior. The patient had no past history and family history of neurofibromatosis. There was no previous trauma, surgical procedure or skin disease at the site. Ultrasonography revealed a well-defined, oval, solid and hypoechoic mass (4.88 cm × 3.9 cm × 3.55 cm). No blood flow was noted in the lesion during color Doppler survey Electro diagnostic studies showed a prolonged distal latency and decreased amplitude of tibial compound motor action potential with proximal stimulation. Predominant slower motor nerve conduction velocity was noted at the segment of the popliteal fossa. There was no sensory nerve action potential elicited at stimulation of the right sural nerve. An electromyogram (EMG) study revealed increased polyphasic waves in the right lateral gastrocnemius, whereas no abnormal waveform was noted in the right tibialis anterior (Fig. 2 a, b).

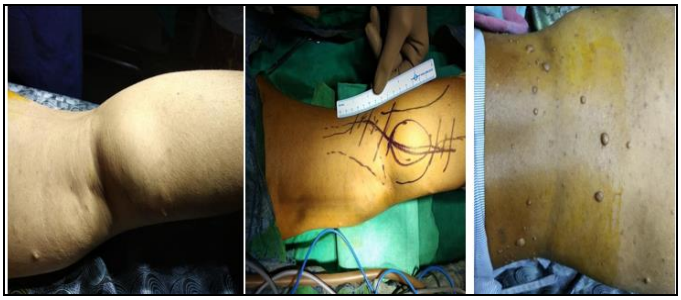

Fig 1: A: Swelling in popliteal fossa **B:** preoperative **C:** Neurofibrometosis

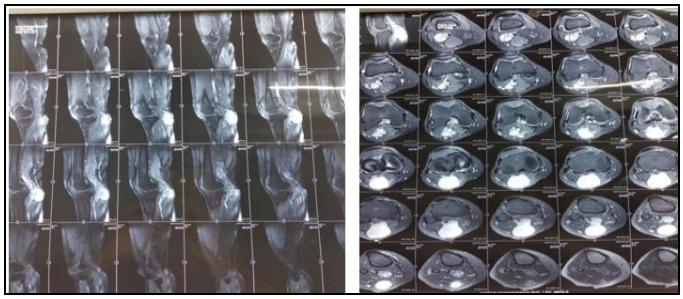

Fig 2: A: MRI Vertical **B:** Cross section MRI

These findings were indicative of an entrapment lesion of the right tibial nerve at the knee level. Magnetic resonance imaging (MRI) showed a well-defined nodule of approximately 7cm in diameter at the right popliteal fossa just behind the popliteal artery and vein. An echo-guided biopsy was performed and a microscopically pathological examination showed a spindle proliferative lesion in the fibromyxoid background, which confirmed the diagnosis of Neurofibroma/Schwannoma. The patient underwent surgical excision of the mass. Post-operative status - complete relief of pain and hypoesthesia. Neurovascular status normal (Fig. 3a, 3b)

Fig 3: A: Speciman after removal **B:** In bowel specimen

The post-operative findings revealed a tumor mass enveloped by the tibial nerve (Fig.4, 5)

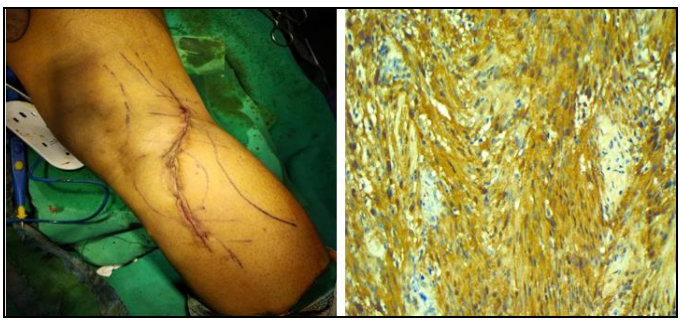

Fig 4: Post-Operative Scar

Fig 5: Histo-Pathology

A surgical pathological examination also confirmed the preoperative diagnosis. No complications were found and the pain was alleviated after excision of the Neurofibroma. Discussion Two genes, NF1 and NF2, are related to neurofibromatosis. Neurofibromatosis type 1 is caused by a mutation of the NF1 gene on chromosome 17q

Discussion

In addition to arising from Schwann cells, neurofibromas incorporate all sorts of cells and structural elements such as perineural cells, mast cells, pericytes, endothelial cells and smooth muscle cells [6, 7]. Multiple cutaneous neurofibromas are common in those with neurofibromatosis type 1, and there are usually multiple lesions over the trunk, face or limbs [1]. Solitary neurofibroma, superficial or deep localization, is less common [1]. It can occur in any nerve of the body with variable symptoms, e.g. asymptomatic, progressive pain, weakness, tingling, or numbness. A palpable mass with a solid texture is the initial finding during physical examination. Tapping on the mass may cause electric pain to shoot down the extremities. Ultrasonography plays an important role in the initial examination of soft tissue mass. Solitary neurofibroma is often hypoechoic and uncompressed, with or without intrinsic blood flow [8]. A traceable nerve can be found, which confirms the tissue origin. However, there is considerable overlap between ultrasonographic appearances of neurofibromas and neurilemmomas [9]. Ultrasonography cannot reliably distinguish between them [8]. Physician can make a tentative diagnosis based on the history, physical examination and initial ultrasonography. However, definitive diagnosis is obtained by MRI and surgical biopsy. Our patient presented with a palpable mass over the right popliteal fossa with tenderness on palpation and sensation was altered along the distribution of the medial sural cutaneous nerve. The texture of the mass felt hard and solid. Since normal cutaneous figuration without coagulopathy and traumatic history were noted, a hematoma was thought to be unlikely. In addition to no pulsation or bruit on auscultation, there was no signal in the Doppler echogram to rule out the possibility of an angiomatous tumor. An abscess or infected popliteal cyst was an unlikely possibility since the patient did not show any evidence of systemic or local signs of infection. Ultrasonography revealed a well-defined and echo-poor mass with fixation at a deep location. An intranervous mucoid cyst was another consideration in addition to a soft tissue tumor. Because of the poor echogenicity of the mass and structural homogeneity with hard texture, the possibility of a soft tissue tumor was highly suspected. Tracing its origin carefully, the mass seemed to lie on the pathway of the tibial nerve. A neurofibroma or neurilemmomas was the tentative impression. The best method of investigation was musculoskeletal ultrasonography due to its high lesion detection rate and diagnostic accuracy, combined with its low cost, wide availability, and ease of use [10, 11]. MRI offers another useful route in studying features of soft tissue masses. Deep neurofibromas have unique MRI characteristics, which are usually homogeneous or heterogeneous with a target pattern. The target-like MRI appearance of deep neurofibromas reflects their histological composition and their location along the course of major nerves [12]. In our patient, MRI revealed the location of the tumor enveloped by the tibial nerve, which was adjacent but distinct to the popliteal vessels. In comparison with MRI, however, ultrasonography turned out to be the better choice for diagnosing the soft tissue tumor because it was performed in real time and was low cost. However, the MRI study gave us better information about the location of the lesion

relative to the adjacent structures. Nerve conduction velocity and EMG studies can also provide information on the localization and nature of a lesion (demyelinating and/or axonal one). These studies were helpful in the assessment of the prognosis and consequential follow-up of the recovery after surgical excision. According to the preoperative electrodiagnostic studies, it was likely that the lesion involved axonal loss and demyelination, consistent with a drop in tibial compound motor action potential amplitude and decreased motor nerve conduction velocity. EMG revealed abnormal musculature of the gastrocnemius but the tibialis anterior was normal, which suggested that the lesion was along the tibial nerve. Neurofibromas are commonly treated by surgical removal. Cutaneous neurofibromas are not usually removed unless they are predominantly painful, with neurologic deficit, or due to cosmetic consideration [13]. In conclusion, we have presented a case with a neurofibroma in the right popliteal fossa, which is a rare location. Solitary neurofibroma can be diagnosed by the history, physical examination and ultrasonography. The differential diagnoses of a solitary mass include hematoma, abscess, intravenous mucoid cyst, neurilemmoma and neurofibroma. Nerve conduction velocity and EMG are helpful in the assessment of prognosis and postremoval follow-up if neurologic defects are noted.

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