



ISSN (P): 2521-3466
ISSN (E): 2521-3474
© Clinical Orthopaedics
www.orthoresearchjournal.com
2018; 2(3): 01-08
Received: 01-05-2018
Accepted: 05-06-2018

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A report of 4 cases, on a rare entity of schwannoma in limbs i.e, outside the central nervous system

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Abstract

Schwannomas are lobulated, encapsulated tumours that arise from the neurilemmal cells in nerve sheaths. Schwannomas are commonly seen in the central nervous system. Its occurrence in the peripheral nervous system is rare. Hence we decided to report our series of 4 cases of schwannomas in limbs. I have excised all of them at different institution and confirmed our diagnoses with histopathology report. I am here with describing some of the cases.

Keywords Schwannoma, biceps tendon sheath, peripheral nervous system

Introduction

Schwannomas are derived from Schwann cells of neuroectoderm. Their function is to form the myelin sheath of nerves in the peripheral nervous system, which insulates the nerve and facilitates the transmission of an impulse. Schwannoma is a benign encapsulated slow growing tumour^[9, 10]. Unlike neurofibromatosis schwannomas do not traverse through the nerve but remain in the sheath lying on top of the nerve. They have a low risk of metastasis^[11]. Schwannomas are most common in patients in second decade and have no gender or racial predilection^[12]. They present with no symptoms, mild symptoms or severe symptoms mostly affecting nerves^[13].

Malignant transformation of a schwannoma (neurilemoma) is an exceedingly rare event. Seven acceptable cases were found in the literature. Analysis of the nine cases of schwannoma with malignant transformation showed no sex predilection, but revealed a tumour differing significantly from conventional malignant peripheral nerve sheath tumours. Rather, the malignant component was commonly purely epithelioid^[14, 15]. The prognosis for patients with schwannomas undergoing malignant change is poor. No significant signs of malignant transformation were seen in our case at follow-up of six months.

Method and materials

Three patients with swelling, pain and paresthesias depending upon the site of tumor and numbness over adjacent and path of nerve, in the age group of 16- 52 years reported in department of Orthopedics at various hospital, where I have been working, from April 2011 to December 2016. There were 2 males and 2 female patient. There was history of pain, swelling over distal part of the tumor with gradual increase in size of swelling. There is also history of paresthesias, burning sensations and Tinel's sign over distal part of the nerve, here are individual case; at different institutions.

Case no 1

A 20-year-old male presented with a 2-month history of palpable mass at the posterior aspect of the knee without any history of trauma. He complained of a growing mass at the posterior aspect of the distal thigh, just below the knee joint. The mass was rubbery hard and it was tender on rolling with finger and Tinel's sign was negative. There was no gross evidence of motor weakness, sensory loss of the lesion.

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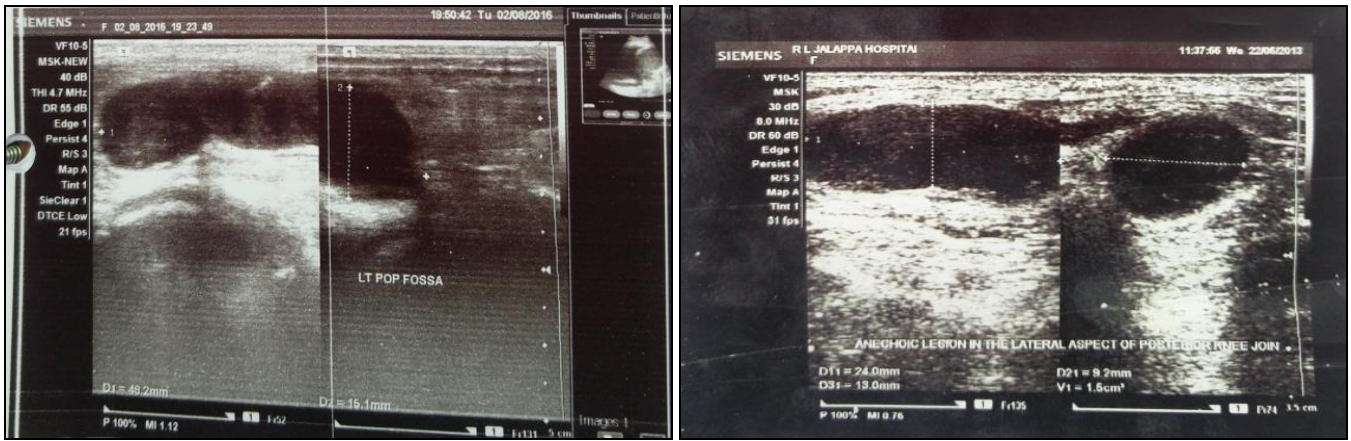


Fig 1: Ultrasound report: There is well defined lobulated echogenic lesion noted in posterior-lateral aspect of right knee in intramuscular plane with few internal septations and mild internal vascularity in colour Doppler.

Diagnostic imaging included plain radiograph and magnetic resonance images (MRI) of the patient's right knee. Routine radiography showed a subtle increase in soft tissue density on the lateral view of the knee. MRI demonstrated 4.6×1.7×1.8 mm (craniocaudal-anterioposterior-transverse) soft tissue mass located just deep into lateral head of gastrocnemius and biceps

tendon muscle. There is another cystic lesion medial to popliteal vein in inter muscular plane measuring 16-5.5-7.0mm (craniocaudal-anterioposterior-transverse). The lesion displayed a well-demarcated oval shape, had lower signal intensity on T1-weighted image, and displayed heterogeneous high signal intensity on T2-weighted images



Fig 2: Magnetic resonance images of the knee. (a): axial T1- and T2-weighted images showing location of the lesion; (b): coronal and sagittal T2-weighted image sequences with high signal intensity on peripheral rim with intermediate signal intensity at the center of the mass showing 'target sign'.

A 8 cm over-the-top incision was made to posterolateral aspect of the knee. The tumor was located at the tendon of the biceps femoris muscle tendon sheath. The mass seemed adherent to tendon and subcutaneous tissue but was not arising from either. The nodule was firm, glistening in appearance, kidney shape and bluish to white in color measuring approximately 4 cm × 6 cm in

size. It was well capsulated without adhesion to the surrounding soft tissue. The lesion was 'shelled out' or enucleated from the tendon sheath of the biceps femoris muscle (Fig. 3). Macroscopically speaking, there were no connections with nerve endings or nerve branches or main nerves. Postoperatively, there was no loss of sensitivity or muscle power.





Fig 3: Intraoperative picture showing the relation of the mass to the musculo-tendinous junction of biceps femoris muscle tendon sheath. Removed mass. Cut section of Mass.

Intraoperative picture showing the relation of the mass to the musculotendinous junction of biceps femoris muscle tendon sheath.

The histological review demonstrated findings consistent with schwannoma, as well as histochemical staining consistent with the presence of the S-100 protein

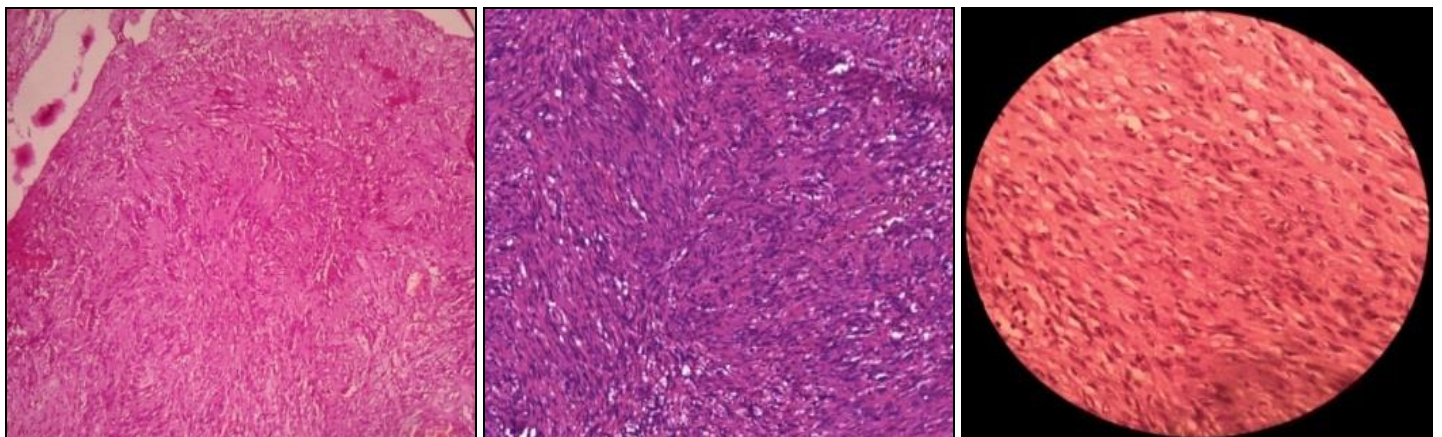


Fig 4: Microphotography of the neurilemoma. (A). Dense Antoni A and loose Antoni B areas (hematoxylin and eosin $\times 100$). (B). Positive S100 protein staining (hematoxylin and eosin $\times 200$).

Case 2.

A 30 year old male patient presented to us with complaints of pain over back of knee and calf region since 12 months. Pain aggravated with exertion, walking and relieved on flexing the knee. He gave history of shooting pain on stretching the leg and Tinel's sign positive. Since six months, he was started with symptomatic treatment with analgesics, for which he responded and was able to manage his daily activities. As the pain and numbness worsened later and was not responding to analgesics, patient turned up for follow up. On examination, there was no palpable swelling even on resisted flexion of knee. There was shooting pain on pressing the swelling. He had superficial point tenderness over his lower popliteal region, and Tinel's sign was positive.

An ultrasound scan yielded well circumscribed, well defined hypo echoic lesion measuring 1.8x1cm noted in popliteal fossa on posteromedial aspect in muscle



Fig 5: Ultrasound of Popliteal Region over the area of Tenderness.

He was planned for excisional biopsy. Preoperatively tender point was marked and dissection in muscular plane over it has yielded a pearly white spherical swelling in the superficial cutaneous nerve sheath [Figure 6]. Marginal resection was done and specimen sent for histopathology [Figure 7]. Histopathology report confirmed schwannoma [Figure x]



Fig 6: Dissection in Surgical Plane Resected Gross Specimen

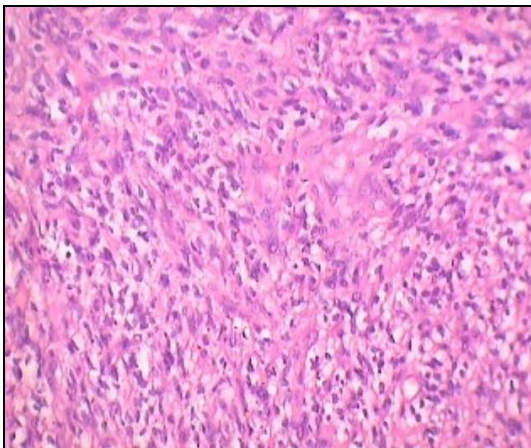


Fig 7: Histopathology Slide

Case 3

A 12-year-old female patient presented to us with complaints of swelling over the anterolateral aspect of right leg lower third region since 1 year (Figure 8). Swelling was insidious in onset, initial size being that of peanut which gradually progressed to about 5×3×2 cm at the time of presentation. Patient did not give any history of trauma/prick injury. Patient gave history of previous unsuccessful attempt of excising the mass 3 months back. No history of constitutional symptoms and any other similar swellings in the body. On examination proper, swelling extended from 3 cm from tip of lateral malleolus to 8 cm from lateral malleolus. Swelling was ovoid in shape with Small wound over anterior aspect of swelling with serous discharge.

Swelling was firm in consistency with mild tenderness and no local rise of temperature. It was non-reducible and non-transilluminant, immobile in both horizontal and vertical directions. Sensations were diminished over dorsum of right foot. No motor weakness present. Movements at right ankle joint were normal.

Gross examination of the biopsy specimen revealed greyish white bits measuring 4×3×2cm, along with multiple tiny bits altogether measuring 2×1×1cm. cut section shows homogenous grey white nodular areas (Figure 9).

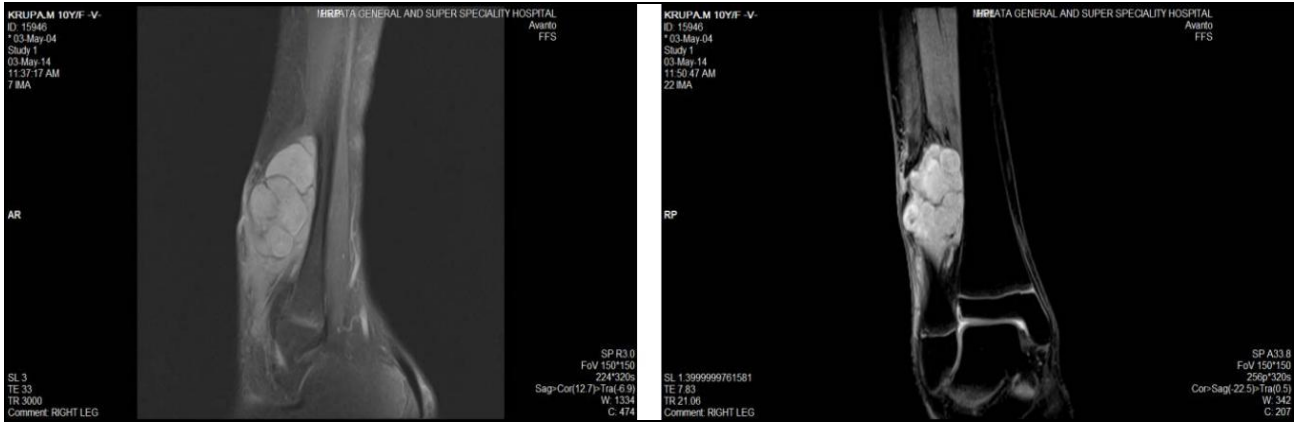
Microscopic examination smear showed Antoni A area composed of spindle shaped Schwann cells arranged in interlacing fascicles without any hemorrhage and necrosis (Figure 10, Figure 11).

A schwannoma of the superficial peroneal nerve is very rare. There were no available data on the incidence of this occurrence. The first report [2] describes a patient presenting with pain at his right fourth toe and at his dorsal fourth web space. This pain was worsened when pressure was applied at the mid-calf region. On surgical exploration, and subsequent histological investigation, a benign schwannoma of the deep Peroneal nerve was found. Post-operatively his symptoms had completely abated.

The second report [3] describes a patient who had presented with an 8-year history of paraesthesia on the dorsal aspect of her right foot, and a mass in the lateral aspect of her popliteal fossa. A 30-cm long tumour was excised from the lateral peroneal nerve. Subsequently the patient developed a right sided foot drop, and paraesthesia on the anterior and lateral aspect of the right lower leg.



Fig 8: Clinical Picture of swelling over Fibula showing erosion of the lateral cortex lower third of leg



(A) (B)
Fig 9: T1 and T2 images show multi-lobulated swelling over lower third of leg



Fig 10: Resected tumour

brevis muscle, exhibiting intermediate signal intensity on T2 weighted images [Fig. 12] and hyperintense signal on FSEIR image [Fig. 13]. Later on patient was taken up for surgery. Excision of the lesion was performed and subjected to histopathological examination. Histology report described the lesion as a benign schwannoma with no evidence of malignant change [Fig. 14].

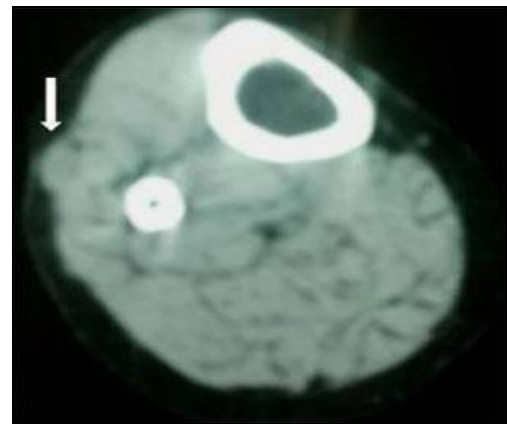


Fig 11: Axial CT scan below the level of fibular neck showing a well-defined round to oval soft tissue density lesion in the peroneal compartment on the lateral aspect of left leg (white arrow).

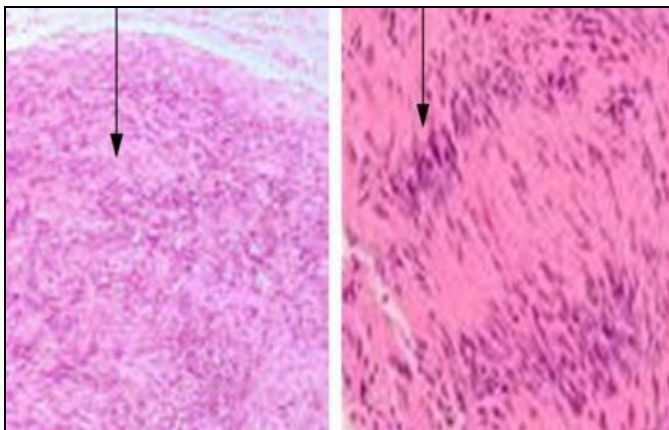


Fig 11: HPE showing Antoni A area composed of spindle shaped cells

Case 4

A 52 years old female presented to us with chief complaints of pain and paraesthesias in the lateral aspect of upper one third of left leg. The pain was moderate in intensity, burning in character and off and on in nature. There was no diurnal variation or radiation of pain. On examination, there was moderate tenderness in the same region. There was reduced sensation along the lateral aspect of the left leg. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) of the leg were advised for further evaluation. CT revealed a well-defined round to oval soft tissue density lesion in the peroneal compartment on the lateral aspect of left leg below the fibular neck [Fig. x]. MRI revealed a round to oval lesion measuring 9x8 mm below the fibular neck in the peroneal compartment indenting the peroneus

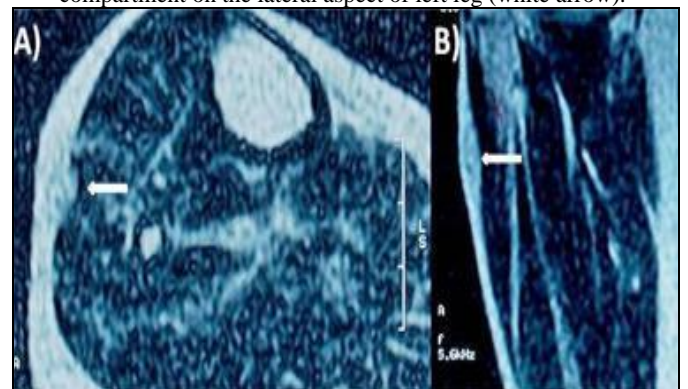


Fig 12: Axial and Coronal T2 weighted MRI images showing a round to oval lesion intermediate signal intensity lesion below the fibular neck in the peroneal compartment indenting the peroneus brevis muscle (white arrows)

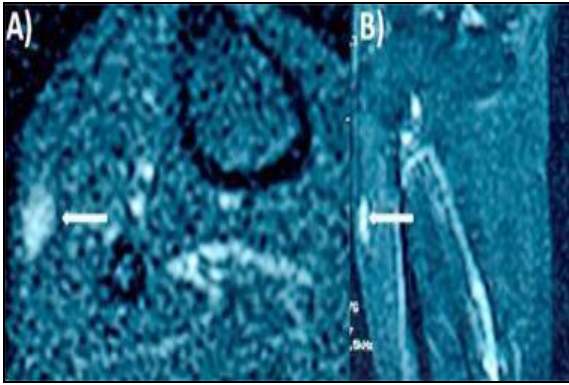


Fig 13: Axial and Coronal FSEIR images, showing a hyperintense intensity lesion indenting the peroneus brevis muscle in the peroneal compartment.

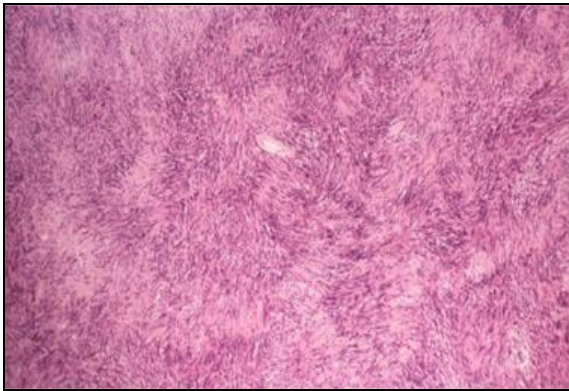


Fig 14: Histological Slide signal (haematoxylin-eosin low power magnification) showing areas of moderate to high cellularity and Scant stromal matrix comprising of elongated cells with cytoplasmic processes arranged in fascicles along with less densely cellular areas consisting of loose meshwork of cells confirming the diagnosis of schwannoma

Results

All the three patients had gradual improvement in symptoms of compression neuropathy over period of 4 weeks and complete resolution of symptoms over 3 months. There was no case of fracture, ankle joint instability, sural nerve or peroneal nerve injury and infection.

Discussion

Case 1

Schwannoma has a predilection to the lower limbs of the body particularly the thighs, buttocks, groin, leg, and retroperitoneum [5]. On clinical examination of the calf and thigh, a major difficulty is when a swelling is deep to the fascia near to nerve sheath, making it difficult to appreciate as a lump [6]. A general ultrasound with clinical symptoms would provide useful additional information. It is non-invasive and has an important role in differentiating a cystic and a solid swelling and its size, thereby providing relevant information for further management [7].

Schwannomas are usually well differentiated tumors with non-metastatic potential, especially if they are located in the extremities. Despite the huge size that these tumors can reach, wide excision decreases local recurrence rate to almost zero.

Case 2

Schwannomas or neurilemmomas are lobulated, encapsulated tumours that arise from the neurilemmal cells in nerve sheaths [4]. Schwannomas are derived from schwann cells of the

neuroectoderm. Their impulsefunction is to form the myelin sheath of nerves in the peripheral nervous system, which insulates the nerve and facilitates the transmission, also categorized with a neurinoma, neurilemmoma, or neurofibroma, the schwannoma is a benign encapsulated slow growing tumour [7-8]. Unlike neurofibromas, schwannomas do not traverse through the nerve but remain in the sheath lying on top of the nerve. They have a low risk of metastasizing. Schwannomas were found to have some transmission types that were autosomal dominant [8]. Schwannomas are most common in patients in the second through the fifth decades of life and have no gender or racial predilection [7].

Schwannomas can present with no symptoms, mild symptoms or severe symptoms mostly affecting the nerves. Most lesions are solitary and present as a slowly growing painless soft-tissue mass. Symptoms are unusual, unless the mass has become large enough to compress the adjacent nerve. Infrequently, these tumours can be associated with Neurofibromatosis 1, and in such cases, they are invariably plexiform or multiple lesions. The first case of a solitary schwannoma was discussed by Liebau, who stated that schwannomas should be looked for in all cases where patients present with pain, paresthesia of leg and foot, especially if all other injury has been excluded [9].

CT appearance of schwannomas has been described as a well circumscribed, homogeneous mass of soft tissue density. MRI is especially useful in identifying the exact location and size of the tumour. Schwannomas have isointense signal relative to skeletal muscle on T1-weighted images and increased slightly heterogeneous signal intensity on T2-weighted images [5-6]. On FSEIR images, they have bright signal intensity. Degeneration and cystic cavitations are common in schwannomas. "Ancient" schwannomas refer to long-standing lesions with advanced degeneration exhibiting calcification, hyalinization, and cystic cavitation, findings that can be identified on imaging [10]. Pathologically, schwannomas are fusiform masses that are eccentrically located in relation to the involved nerve and are contained within a capsule, the epineurium. They are composed primarily of Schwann's cells. Surgical excision can usually spare the parent nerve because the schwannoma is generally separable from the underlying nerve fibres.

In conclusion, superficial peroneal nerve schwannomas are a very rare solitary nerve sheath tumours. They should always be considered as a differential diagnosis when tarsal tunnel syndrome, neuromas, nerve entrapment or radiculopathy is suspected. Schwannomas found in the proximal aspect of the lower extremity can also cause distal symptoms or injury, so this must also be considered, especially if the previous differentials have been ruled out. Early diagnosis can prevent permanent nerve damage, soft tissue or bony deformity. Thus in the differential diagnosis of non-traumatic and non-arthritis pain of lower leg and foot, benign tumours especially schwannomas of the peroneal nerves should always be considered.

Case 3

A schwannoma of the superficial peroneal nerve is very rare. There were no available data on the incidence of this occurrence. The first report [11] describes a patient presenting with pain at his right fourth toe and at his dorsal fourth web space. This pain was worsened when pressure was applied at the mid-calf region. On surgical exploration, and subsequent histological investigation, a benign schwannoma of the deep peroneal nerve was found. Post-operatively his symptoms had completely abated.

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foot, and a mass in the lateral aspect of her popliteal fossa. A 30-cm long tumour was excised from the lateral peroneal nerve. Subsequently the patient developed a right sided foot drop, and paraes-thesia anterior and lateral on the right lower leg.

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Malignant transformation of a schwannoma (neurilemoma) is an exceedingly rare event. Seven acceptable cases were found in the literature. Analysis of the nine cases of schwannoma with malignant transformation showed no sex predilection, but revealed a tumour differing significantly from conventional malignant peripher-al nerve sheath tumours. Rather, the malignant component was commonly purely epithelioid [18, 19]. The prognosis for patients with schwannomas undergoing malignant change is poor. No significant signs of malignant transformation were seen in our case at follow-up of six months.

Case 4

This location of schwannoma which arose at the paratenon of the Biceps Femoris Muscle Tendon Sheath, is extremely rare. Only two such cases that developed around the ankle joint have been reported in the literature [20 liebau] and [21 jack]. This is the report of schwannoma occurring at the paratenon of Biceps Femoris Muscle sheath without any neurologic symptoms, which should be included in the differential diagnosis of a lump in the posterior aspect of the knee.

Liebau *et al.* [20] reported a schwannoma arising at the flexor digitorum longus muscle that was located between the tendon mirrors of the two muscles, which evoked pain and parathesias. Jack *et al.* [21] reported a case of multiple schwannomas arising at the medial aspect of Achilles tendon, which caused pain and positive Tinel's sign.

Schwannomas can develop anywhere in the body which are most prevalent in major nerve trunk or peripheral nerve at the upper extremities and lower extremities. Numerous previous reports of schwannoma localized to the region around the knee were schwannoma arising from peroneal nerve near the fibular head [22 kim], [23 knight] and [23 nawabi].

The clinical features of schwannoma in the limbs usually present as a pain, mobile, and Tinel's sign, in which percussion over the lump induces painful paresthesia, are common [33 kim] and [22 knight]. However, our case had tenderness on deep palpation, no paresthesia, or Tinel's sign, but only palpable lump at the posterior aspect of the knee.

However, in our case, mass was located in the tendon/paratenon of the biceps femoris muscle tendon sheath with tenderness, on deep palpation, no paresthesia, and Tinel's sign, but only palpable lump. To our knowledge, tendon itself does not contain nerve fibers [324 benjamin]. We made an assumption that the mass developed from Golgi tendon organ in musculo-tendinous junction or proprioceptive neural structures on the paratenon since there was no connection of any nerve fibers.

Intraoperative gross findings of schwannoma characteristically

form an eccentric, oval shaped, less than 3 cm in diameter, with the attenuated nerve bundle of the parent nerve stretched and displaced over the mass. However, no connections to nerve fibers or nerve endings were found in the case presented in this report.

Histologically, the neoplasm is characterized by encapsulation, and is composed of Antoni A and B cells and Verocay bodies [33 kim]. In the case described in this report, histological evaluation was done with conventional hematoxylin and eosin staining, and by means of immunostaining for the S100 protein to confirm the diagnosis (Fig. 14) [25 weiss] and [26 Fletcher].

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