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Case Report

A Fatty Spectacle – A Rare Case of Co-occurance of Tibialis Anterior Tendon Sheath Lipoma and Fibrolipomatous Hamartoma of Superficial Peroneal Nerve

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Abstract

Fibrolipomatous hamartoma of nerve is a rare occurrence with only a few reported cases in literature, mostly described in the median nerve. Tendon sheath lipoma is an even rarer entity, mostly described in hands and wrist, with tibialis anterior tendon sheath lipoma being nearly unreported. In the following case report, we present a serendipitous concurrence of these two rare pathologies in one individual without any familial or syndromic association. To the best of our knowledge, such a presentation has not been reported so far.

Key words: Fibrolipomatous hamartoma, Musculoskeletal, Synovial lipoma, Tendon sheath lipoma

CASE SUMMARY

A 28-year-old male presented with multiple long standing, painless, solid swellings in the left distal leg, ankle, and foot region, involving dorsum of foot, the lateral malleolus and anterolateral distal leg. There was no history of prior trauma to the region, no functional disability due to swellings, and no significant family history. On examination, no discoloration, prominent vessels, or pus points were seen. Elongated swelling on anterolateral aspect of leg only moved in direction perpendicular to shin of tibia. All the swellings were non-tender.

On ultrasound, elongated swelling on anterolateral aspect of leg and foot appeared hyperechoic with interspersed hypoechoic linear bands. Incidentally, hyperechoic lesion was seen within tibialis anterior tendon sheath, running along the tendon. Few well-defined homogeneous hyperechoic subcutaneous swellings showing reticulations, representing subcutaneous lipomas were also seen around the ankle joint [Figures 1 and 2].

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Magnetic resonance imaging (MRI) was performed which reinforced the ultrasound findings. Axial, coronal, and sagittal images showed fat signal intensity lesion within tibialis anterior tendon sheath – suggestive of tibialis anterior tendon sheath lipoma. The superficial peroneal nerve running lateral and superficial to tibialis anterior tendon was thickened and showed classical "coaxial cable" and "spaghetti appearance" which is suggestive of fibrolipomatous hamartoma. Few subcutaneous lipomas were also noted [Figures 3 and 4].

DISCUSSION

Lipomas are proliferation of adipose tissue usually surrounded by a fibrous capsule and are the most common benign mesenchymal neoplasms. These are just the tip of the iceberg in a spectrum of lipomatous lesions. Benign lipomatous lesions of the soft tissue can be classified into nine distinct categories, which are lipoma, lipomatosis, fibrolipomatous hamartoma of the nerve, lipoblastoma or lipoblastomatosis, angiolipoma, myolipoma, chondroid lipoma, spindle cell lipoma or pleomorphic lipoma, and hibernoma.[1] Benign lipomatous lesions affecting the

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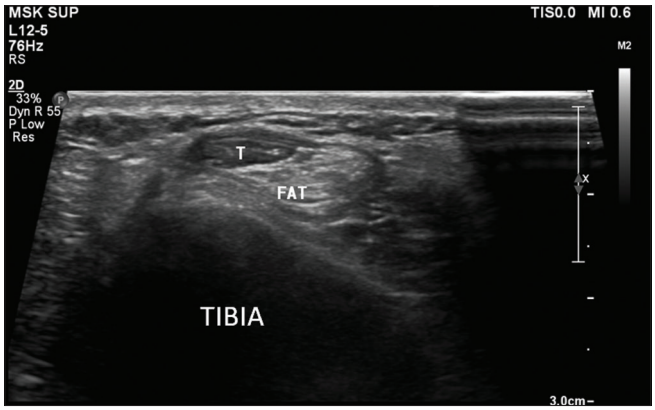

Figure 1: Ultrasound image perpendicular to shin of tibia showing tibialis anterior tendon sheath with hypoechoic tendon and echogenic fat inside the tendon sheath

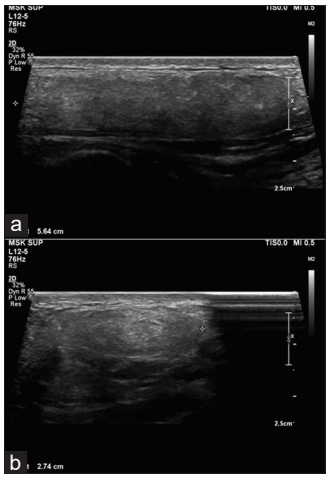

Figure 2: Longitudinal (a) and transverse (b) ultrasound images of elongated chord-like hyperechoic lesion on anterolateral aspect of leg, ankle, and foot showing few hypoehoic linear areas

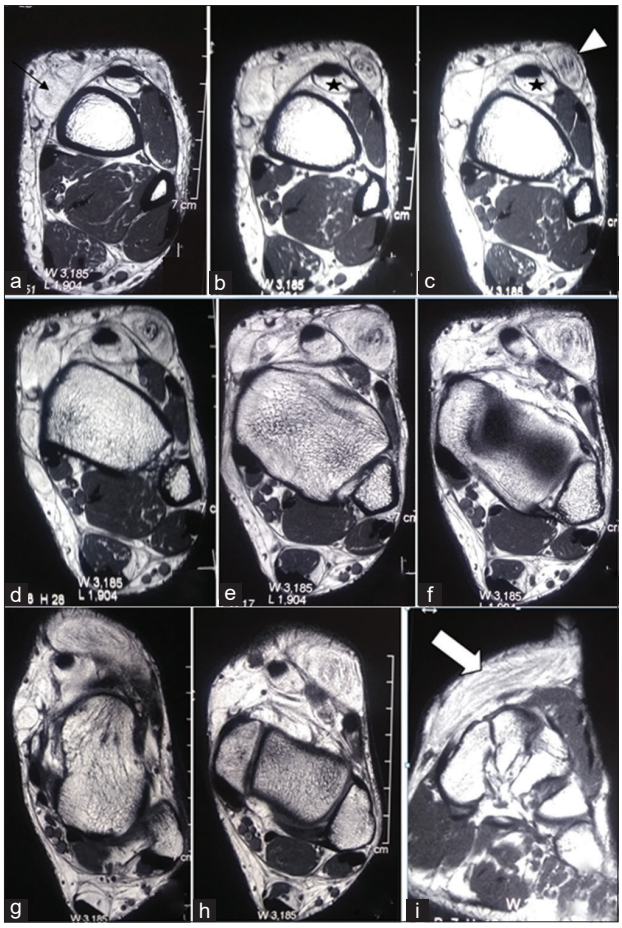

Figure 3: (a-i): Axial (a-h) and coronal (i) T2-weighted magnetic resonance images of the left ankle joint showing hyperintense signal inside the tibialis anterior tendon sheath (star ★). Adjoining superficial peroneal nerve appears thickened and shows "coaxial cable" (arrowhead) and "spaghetti" appearance (white block arrow). Few subcutaneous lipomas are noted on medial aspect of ankle joint (black arrow)

bone, joint, or tendon sheath are much rarer and include intraosseous lipoma, parosteal lipoma, liposclerosing myxofibrous tumor, synovial lipoma of the joint or tendon sheath, and lipoma arborescens.[2]

Subcutaneous lipomas are very frequently encountered as long-standing asymptomatic swellings on trunk and extremities. On the contrary, deeper soft-tissue lipomas are rare and depending on their location, can cause pressure symptoms – like carpal tunnel syndrome in median nerve fibrolipomatous hamartoma.[3] A case report of tendon sheath lipoma causing De Quervain's tenosynovitis has also been reported.[4]

Like most neoplasms, lipomas can be attributed to have genetic origin. Recent studies have shown a correlation between HMGA2 gene located on chromosome 12q14.3 and lipoma production. Another gene EHD-1, which interacts with insulin-like growth factors is purported to have a role in etiopathogenesis of tendon sheath lipomas.[5] Besides genetic link, another theory suggests that there is a direct positive correlation between trauma to an area and lipoma production. In addition to these, other possible risk factors that may lead to lipomas are obesity, alcohol abuse, liver disease, and diabetes mellitus.[6]

Lipomatous lesions are usually sporadic in occurrence. Multiple lipomatous lesions should prompt further evaluation of inherited conditions such as Proteus syndrome, familial hereditary lipomatosis, Decrum's disease, Madelung's disease, Cowden syndrome, and Gardner syndrome.[7]

Synovial Lipoma

Synovial lipomas are a rare entity and can have two distinct presentations – a focal solid fatty mass in tendon sheath and diffuse proliferating fatty synovial thickening, called "lipoma arborescence" typically seen in knee joint.[8]

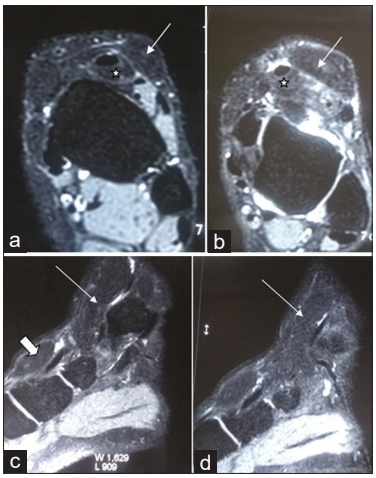

Figure 4: Axial (a and b) and sagittal (c and d) fat-saturated magnetic resonance images of the left ankle joint showing suppression of signal in all the previously described lesions, suggesting fat component. The hyperintense lesion in tibialis anterior tendon sheath shows loss of signal on fat saturation which is suggestive of tendon sheath lipoma (star ☆). Loss of signal is also seen in thickened superficial peroneal nerve (arrow) suggesting fatty infiltration. Subcutaneous swelling also shows loss of signal on fat-saturated images verifying it's a lipoma (black arrow)

Due to their asymptomatic nature and infrequent occurrence, lipomas of tendon sheath can go unreported. True discreet intra-articular lipoma is quite rare and reported more frequently in hand and wrist and infrequently in foot or ankle.[9] These are discrete lipomatous masses, with imaging features similar to those of a superficial or deep lipoma.

On ultrasound, these can be seen along superficial tendons as hyperechoic structures showing few reticulations similar to surrounding fat and no internal vascularity on color Doppler. On computed tomography images, these can be identified by their fat attenuation and appear hypodense to muscles and tendons. They follow fat signal intensity on all MRI sequences, appearing hyperintense on T1, T2, and PD sequences and hypointense on fat-saturated images. Rarely, they may show fluid-like signal intensity which has been attributed to mucoid degeneration.[10]

Lipoma arborescence is the other presentation of synovial lipoma. It is a benign collection of fat replacing the subsynovial layer, resulting in diffuse villonodular proliferation in the synovium that has a frond-like architecture. It affects joints more commonly rather than tendon sheath and is frequently associated with synovial effusion. Cases of extra-articular lipoma arborescens involving peroneal and flexor ankle tendon sheath, subdeltoid bursa, extensor tendon sheath of the hand, and biceps tendon sheath have been reported. Unlike tendon sheath lipoma which is well-defined lesion, lipoma arborescence involving tendon sheath has frond-like architecture.[11-15]

Asymptomatic tendon sheath lipoma can be managed conservatively. Treatment of symptomatic lipoma is surgical excision.[16]

Fibrolipomatous Hamartoma

Fibrolipomatous hamartoma of nerve, as the name implies, is benign hamartomatous deposition of adipose tissue inside a peripheral nerve. The adipose tissue is deposited in between perineural fibers causing expansion of nerve and giving it classical "coaxial cable" appearance on axial sections and "spaghetti appearance" on coronal and sagittal sections.[17]

On MRI, the affected nerve appears thickened and shows discrete T1, T2 hypointense nerve fascicles surrounded by hyperintense adipose tissue. On fat suppression and inversion recovery sequences, the fat signal is suppressed. On ultrasound, the affected nerve appears echogenic and shows hypoechoic discrete fascicles corresponding to MRI appearance.[18]

Median nerve is the most common nerve to be affected. Two-thirds of affected nerves are associated with macrodystrophia lipomatosa.[19] Motor nerves are more frequently involved than sensory nerves. Depending on location, this lesion may be asymptomatic or cause compression neuropathies like carpal tunnel syndrome.[20]

Although mostly described as individual or isolated nerve involvements, rarely, fibrolipomatous hamartoma can be diffuse and involve nerve plexuses such as the brachial plexus or lumbosacral plexus and its branches. Such extensive fibrolipomatous hamartomas can be differentiated from plexiform neurofibromas by the relatively well-defined appearance of affected nerves which are encased in epineurium and maintain their shape as opposed to plexiform neurofibromas which are large irregular masses and do not show signal suppress on fat-saturated images. Association of plexiform neurofibroma with type 1 neurofibromatosis and fibrolipomatous hamartoma with macrodystrophia lipomatosa can also aid the differentiation. Signal loss on fat-saturated images also differentiates fibrolipomatous hamartoma from hereditary hypertrophic interstitial neuritis.[21]

Due to the fact that the lesion is interspersed within the nerve fibers, excision is rarely the treatment option and results in sensory and motor deficits. Management is focused on symptomatic treatment like nerve decompression.[22]

Table 1: Multiple lipoma syndromes

Lipomatosis syndrome	Salient features
Proteus syndrome	Multiple lipomas, hemangiomas, epidermal naevi, scoliosis, hyperostosis of epiphysis and skull
Decrum's disease	Multiple painful lipomas of trunk and extremities, often with paresthesia of overlying skin
Familial multiple lipomatosis	Autosomal dominant inherited disorder. Typically presents in third decade with hundreds of non-infiltrating lipomas
Madelung's disease	Diffuse, infiltrative, symmetric, painless lipomas involving face, neck, and shoulder
Cowden syndrome	Multiple lipomas, oral papillomas, facial trichilemmomas, hamartomatous polyps of the gastrointestinal tract and breast, endometrial and thyroid carcinomas
Bannayan-Riley-Ruvalcaba syndrome	Multiple lipomas, intestinal hamartomas, genital lentigines, macrocephaly, and mental retardation
Gardner syndrome	Gastrointestinal polyposis, osteomas, multiple lipomas, epidermoid cysts, desmoids tumors

Lipomatosis Syndromes

Multiple lipomas can be a manifestation of various syndromes, salient features of some of these are tabulated in Table 1.[23-31]

CONCLUSION

Although very common in subcutaneous plane, lipomas of deep soft-tissue structures such as tendon sheath and nerves are rare entities. Due to their asymptomatic nature and infrequent occurrence, lipomas of tendon sheath can go unreported. Fibrolipomatous hamartoma of nerve affects motor nerves more than sensory nerves and is associated with macrodystrophia lipomatosa in two-third of cases. As the lesion is interspersed within the nerve fibers, excision is rarely the treatment option and management is focused on symptomatic treatment like nerve decompression. Multiple lipomatous lesions should prompt further evaluation for inherited and syndromic conditions. The characteristic radiological appearance usually obviates the need for histopathological confirmation by biopsy. Although two rare forms of lipoma – tendon sheath lipoma and fibrolipomatous hamartoma, both were present in this case along with few subcutaneous lipomas, there was no familial or syndromic correlation. To the best of our knowledge, such a presentation has not yet been reported.

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