Case Report

Arthroscopic Excision of Angio-Fibro-Lipomatous Hamartoma of the Knee: A Rare Case Report

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Abstract

Angio-fibro-lipomatous hamartoma is a benign adipose tissue tumor very rarely seen in musculoskeletal distribution, and its incidence in the knee joint has never been reported. The patient in our case presented with knee pain of 2 years' duration, following blunt trauma. Preoperatively, veno-lymphatic malformation/hemangioma was considered as the diagnosis. Only after arthroscopic excision biopsy, histopathological examination, retrospective radiological analysis, and a review of literature, we were able to diagnose this rare condition. The histopathological picture of this benign adipose tissue tumor contained a mixture of mature adipose tissue and fibrous and vascular tissues. Here, in this case report, we discuss about PTEN gene causing PTEN hamartoma of soft tissue and angiolipoma presentations and its variants.

Keywords: *Angio-fibro-lipomatous hamartoma, angiolipoma, PTEN hamartoma of soft tissue*

Introduction

Intraarticular angiolipomas of the knee can cause knee pain and are uncommon benign soft-tissue tumors that typically arise in the subcutaneous tissues of young patients.[1] Furthermore, the unusual, benign soft-tissue lesions with a characteristic histopathology (admixture of connective tissue elements – primarily fat, fibrous tissue, and abnormal blood vessels) are grouped as[2] PTEN hamartoma of soft tissue (PHOST) following PTEN mutation of the gene. There are only two case reports of intraarticular angiolipoma around the knee which has been reported in the English literature.[3,4] This is the first case report of histological variant of angiolipoma as angio-fibro-lipomatous hamartoma in the knee causing severe pain around the knee. Here, we discuss about a review of literature of PTEN hamartoma, clinical presentation, radiological appearance, and management of our case and other two case reports of this rare benign lesion.

Case Report

A 24-year-old male patient presented with a history of pain in the right knee for 2 years. He had a history of blunt injury by a gas cylinder over the inner aspect of the lower thigh about 2 years back. His pain started after this blunt injury, which was dull aching and occasionally increased in severity on exertion and walking long distance. However, he complained of increased pain for 1 month prior to his consultation in our clinic. He had a second episode of blunt trauma following which pain was present throughout the day which increased on walking even a short distance. He was unable to squat and sit on the floor due to pain. On clinical examination, he was found to have diffuse swelling on the medial aspect of the knee. It was tender to palpation [Figure 1b], and the margins of the swelling could not be demarcated clinically. Swelling turned prominent on flexion of the knee [Figure 1a and c]. The skin over the swelling was normal. The patient was initially evaluated with X-rays and since the clinical diagnosis could not be made, the patient was further evaluated with magnetic resonance imaging (MRI), computed tomography (CT) scan, and ultrasonography (USG) screening. X-ray examination was unremarkable, and MRI was reported to have venolymphatic malformation over the medial femoral condyle with minimal extension into the medial femoral condyle and vastus medialis obliqus. All the other blood parameters such as erythrocyte sedimentation rate, C-reactive protein, and rheumatoid factors

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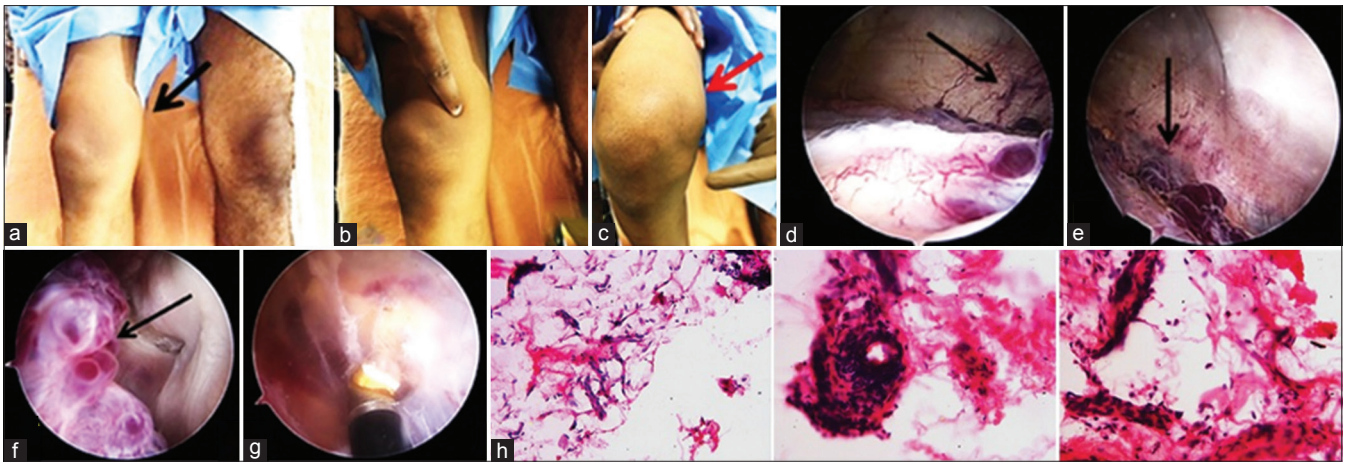

Figure 1: Clinical and arthroscopic picture: (a and b) diffuse swelling and tender to palpate (c) swelling appeared prominent on flexion of the knee, (d) arthroscopic picture – vascular nature of the lesion, (e) arthroscopy demonstrates extension into the medial capsule and vastus medialis, (f) grape-like multilobulated cystic tissue resembling hemangioma, (g) the lesion was debrided with radiofrequency probe over the medial capsule and medial femoral condyle and over the articular surface of the vastus medialis, (h) histological picture – mature adipose tissue, fibroblasts, and thick-walled dilated vessels

were within the normal limits. The patient was taken for diagnostic arthroscopy and excision biopsy. Through standard anteromedial and anterolateral portals, the knee joint was examined; there were no intraarticular bleeding tendencies until the lesion was probed. Multiple, small, dark maroon, grape-like multilobulated cystic tissue [Figure 1d-f and Video 1] was identified in the medial gutter over the medial femoral condylar area and over the medial wall extending into the medial capsule and articular surface of the vastus medialis muscle. Rest of the knee corners were normal, and the lesional area was identified to be highly vascular [Figure 1d] with prominent vessels seen arthroscopically. The tissue was bleeding on probing, biopsy of the lesion was taken, and extension into the bone and vastus medialis area was debrided with shaver and radiofrequency (RF) probe [Figure 1g]. RF uses rates of oscillations (rapid movements/vibrations in the back-and-forth motion) in the range of around 3 kHz to 300 GHz (which corresponds to the frequency of radiowaves, and the alternating currents which carry radio signals). RF usually refers to electrical rather than mechanical oscillations, but mechanical ones do exist.[5] The process of removing all the tissues takes place with the RF probe. The main goal is to perform coblation, which is controlled ablation. Ablation is the removal or cutting of tissue, and this offers the form of cutting or removing tissue by delivering large amounts of energy or heat. First, a plasma layer of reactive particles that disrupts the tissue's organic bonds is applied which rapidly disintegrates the tissue. This allows for large amounts of tissue removal with minimal damage to the surrounding tissue. This plasma layer is created by a current flowing between closely spaced electrodes. It vaporizes a layer of conductive fluid, and then the plasma layer is formed which is very reactive with energized electrons and ions. Next, an ablative layer is established quickly, using high current density, and then coblation occurs.[4-6] Hemostasis was achieved, and wound was closed over a drain. The postoperative period was uneventful, and the patient had very good pain relief after the surgery. He was asymptomatic at the time of final followup of 1 year.

Discussion

Benign adipose tissue tumors can be seen in many conditions. For example, lipomas, lipomatosis, lipoblastoma, lipoblastomatosis, angiolipomas, and myolipomas are considered as benign adipose tissue tumors.[7] Angiolipomas are rare benign tumors with potentiallyno malignant transformation.[8] They have typical histological appearance of mature fatty tissue and sprinkled proliferated vascular components.[9] Angiolipomas comprise 5%–17% of lipomas[7] and generally seen in subcutaneous regions of forearm,[8] triceps surae muscle (hemartoma)[10] and other areas such as intracranial region,[11] neck,[12] colon,[13] and mesentery of the sigmoid colon[14] are reported in the literature. Histological variant of angiolipoma with fibrous hamartoma in knee [Figure 1h] has not been reported and, to our knowledge, this is the first case report. Angiolipomas are rarely seen in knee joint and till date, only two case reports have been published in the English literature. The first case was reported in 2010 by Nishimori *et al*.,[15] where the patient presented with recurrent pain and hemarthrosis followed by arthroscopic excision from posterior compartment of knee (behind posterior cruciate ligament). They reported that angiolipoma of knee usually presents with pain, hemarthrosis, and dysfunction;[13] however, in our case, this vascular lesion did not present with hemarthrosis though his onset of symptoms was from 2 years. On arthroscopic examination, this vascular lesion did not show any spontaneous bleeding tendency until probed. The second case was published by İlyas *et al*.[8] of a 13-year-old boy with a giant-sized multilobular noninfiltrating angiolipoma (completely encapsulated), extending outside the knee joint and causing lateral patellar

dislocation. Here, the author had performed tibial tuberosity transfer to restore the patellofemoral articular alignment in addition to complete resection of tumor mass and repair of medial retinaculum.[3] However, in our case, the lesion was intraarticular with a small portion of the benign mass extending into the medial femoral condyle and vastus medialis obliqus [Figure 2], suggesting the infiltrative nature of the mass. Gonzalez-Crussi *et al*. (1966)[16] proposed classifying angiolipomas into infiltrating and noninfiltrating types.

Radiological appearance in coronal images showed a well-defined lobulated lesion isointense to muscle in T1-weighted image [Figure 2a] and heterogeneously hyperintense on T2-weighted images [Figure 2b] along the medial femoral condyle which is extending to involve the medial vastus muscle as seen in fat-saturated T2-weighted axial image [asterix in Figure 2c]. There is also a small intraosseous component in the adjacent medial condyle which is seen as widened intertrabecular spaces and thickened adjacent trabeculae as seen in axial fat-saturated T2-weighted image and axial CT images [curved arrow in Figure 2d and e]. USG screening showed slow-flow vascular channels within it [arrowhead in Figure 2f]. T2-weighted images also showed hypointense septations suggestive of fibrous component. USG screening also showed hyperechoic areas within the lesion which were of low signal intensity on CT scan, suggesting fat content. In view of fibrous, vascular, and fat contents within the lesion, a benign slow-flow venolymphatic malformation with lipomatous and fibrous components was considered. There was no evidence to suggest intraarticular bleed on imaging. Thus, from these histological and radiological appearances, this is the only case report demonstrating infiltrative and variant nature of angiolipoma [Figure 2]. Kurek *et al*.[2] in their series of 34 patients had found that these lesions did not have a distinct capsule, and they often involve the muscles and subcutaneous tissue simultaneously, surrounding the neurovascular structures. These findings also comply with our case which had infiltration into the medial femoral condyle and vastus medialis muscle.

The etiology of this rare benign adipose tissue tumor is mostly accepted by the authors that angiolipomas may be developed by embryonic sequestration of multipotential mesenchymal cells, and puberty activates this process via hormones.[17] However, this histological variant of the angiolipoma may be grouped under PHOST which could be due to germline mutation in the PTEN tumor suppressor gene[18] on 10q23.3 as described by Kurek *et al*.[2] They have also found that this mutation was associated with rare syndromes such as Bannayan–Riley–Ruvalcaba syndrome and Cowden's syndrome. In our case, the patient did not exhibit any of the features of these syndromes and examination of the patient in search any occult hemangiomas/hamartomas and other benign lesions were futile. While we have termed this lesion as hamartoma, disproportionate growth relative to the location might not be included in the most stringent definition of hamartoma (normal tissue in abnormal location). This case may be

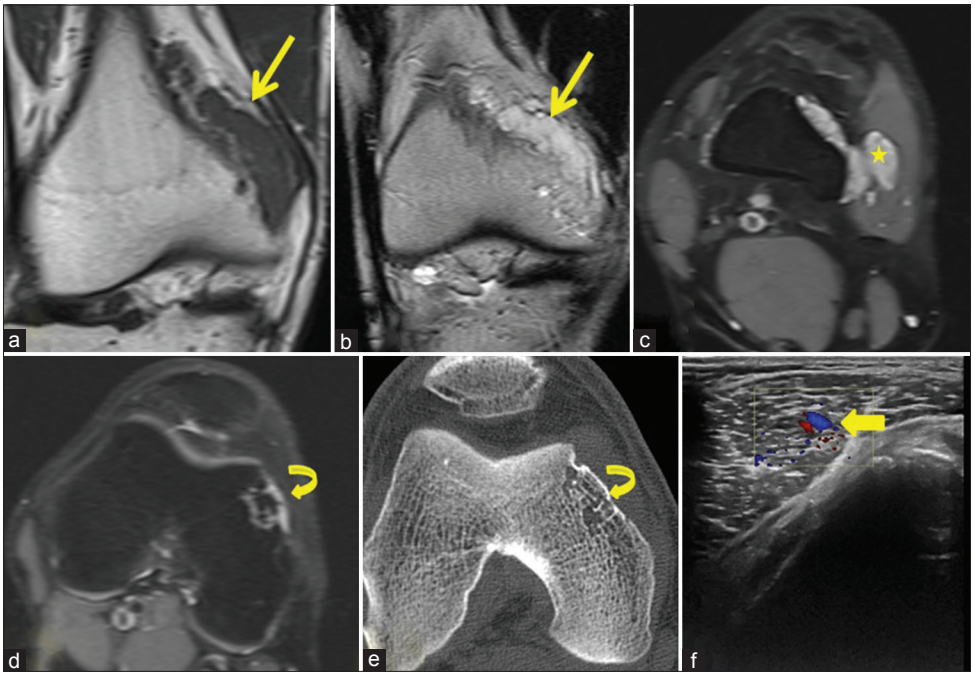

Figure 2: Magnetic resonance imaging, computed tomography, and ultrasonography screening, (a) coronal magnetic resonance imaging shows a well-defined lobulated lesion isointense to muscle in T1-weighted image (arrow in a), (b) heterogeneously hyperintense on T2-weighted images (arrow in b) along the medial femoral condyle which is extending to involve the medial vastus muscle as seen in fat-saturated T2-weighted axial image (asterix in c), (d and e) there is also a small intraosseous component in the adjacent medial condyle which is seen as widened intertrabecular spaces and thickened adjacent trabeculae as seen in axial fat-saturated T2-weighted image and axial computed tomography images (curved arrow in d and e), (f) ultrasonography screening showed slow-flow vascular channels within it (arrowhead in f)

included as a part of PHOST purely on the resemblance with the histological variant of angiolipoma and also the infiltrating nature into the muscle and soft tissue, as reported by other authors.[12,13,17,19] The role of PTEN mutations in the development of PHOST is not fully understood. PTEN affects cell growth and/or migration of the various components of PHOST;[16,19,20] this being a rare lesion, it is difficult to prove its association with PTEN mutation; nevertheless, possibility of the mutation cannot be ruled out as the antecedent blunt trauma could have been incidental and would have just contributed to increase in the severity of the pain. The association of blunt trauma and rare lesion like this case has not been reported yet. and its relationship is inexpedient to prove. After extensive literature search, rare lesions such as hamartomatous lipofibrovascular mesenchymal tumor-like mass are found in the mesentery of the sigmoid colon,[2] but their incidence around the knee has not been reported till date.

Conclusion

A rare lesion like angio-fibro-lipomatous hamartoma may cause severe knee pain, and arthroscopic examination will be useful in identifying the gross appearance and diagnosing rare lesions. Possibility of gene mutations needs to be ruled out, and search of occult lipomas in other parts of body will be helpful in identifying certain rare syndromes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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