



ISSN: 2395-1958
IJOS 2018; 4(1): 463-466
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www.orthopaper.com
Received: 20-11-2017
Accepted: 22-12-2017

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Solitary Extrarenal angiomyolipoma in the knee: A case report and literature review

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DOI: <https://doi.org/10.22271/ortho.2018.v4.i1g.68>

Abstract

Angiomyolipoma is usually a solitary renal tumour and composed of adipose tissue, blood vessels, and smooth muscles. Extrarenal angiomyolipoma is an uncommon disease and commonly found in liver. Angiomyolipoma in the musculoskeletal tissue particularly in the subcutaneous region of the knee is very rare. Histopathological section of angiomyolipoma shows an admixture of small to medium-sized thick-walled blood vessels, smooth muscle cells and adipose tissue which include different types of mature fat cells. The treatment of AML is wide resection, which is usually curative. We describe a case of Angiomyolipoma (AML) in the subcutaneous tissue of the knee joint of a 70 years old male with MRI and histological features of the tumour. There is an only one previous report of this type of case in the literature. To the best of our knowledge, this is the second case of a solitary angiomyolipoma in the subcutaneous tissue of the knee joint area.

Keywords: Angiomyolipoma, subcutaneous tissue, knee

Introduction

Angiomyolipoma (AML) is a mesenchymal neoplasm composed of variable proportions of blood vessels, smooth muscle, and adipose tissue. The most common site is kidney. The majority of AMLs are sporadically occurring solitary renal tumours (80% of cases) and only occasionally it manifests as part of tuberous sclerosis complex (TSC). Extrarenal angiomyolipoma is an uncommon disease and usually found in liver [1]. It is also found in various other sites; however, angiomyolipoma in the musculoskeletal location is very rare. Few cases have been reported in the acral soft tissues [2, 3, 4], skeletal muscle [5] and bone [6]. We describe a case of Angiomyolipoma (AML) in the subcutaneous tissue of the knee of a 70 years old male with clinical, histological features and MRI findings of the tumour. To the best of our knowledge, there are no previous reports of this type of tumour in our country. There is an only one previous report of this type of case in the literature [2].

Case Report

A 70 years old male came to my clinic with a swelling in the medial aspect of his right knee. He noticed a small lump in the postero-medial aspect of the right knee 7 years back. The lump was gradually increased in size with mild dull aching pain. He had no history of trauma and infection. He did not take any treatment for his ailment.

On examination, there was a 5 × 3 cm ovoid lump on the medial aspect of the right knee, at the level of joint line (Figure 1). It was smooth, firm, not tender and not fixed to overlying skin and underlying structures. The lump was mobile and trans-illumination test was negative. Regional lymph nodes were not palpable and neurovascular status of the lower limb was intact.

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Fig 1: An ovoid lump on the medial aspect of the right knee.



Fig 2: Plain X-ray of the right knee showing a soft tissue swelling on the medial side of the knee.

Laboratory investigations were unremarkable. Radiographs of the knee along with magnetic resonance imaging (MRI) of knee were obtained. Radiograph of the knee was unremarkable except mild degenerative changes and a soft tissue swelling on the medial side (Figure 2).

Coronal and axial (Figure 3 and 4) magnetic resonance (MR) images of knee revealed a subcutaneous, encapsulated, well circumscribed, well defined mass with mixed soft tissue densities.



Fig 3: MRI (Coronal section) showing an ovoid subcutaneous mass on the medial side of the knee.

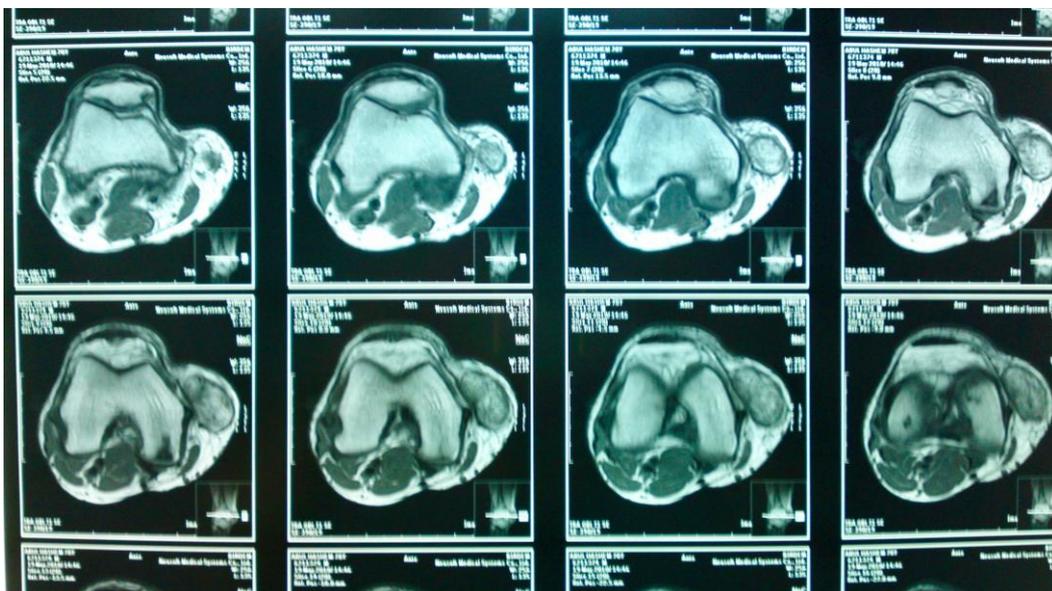


Fig 3: MRI (Axial section) shows a subcutaneous, well circumscribed mass.

The mass was removed by local wide resection under spinal anesthesia. It was subcutaneous and encapsulated. There were no adhesions to the surrounding structures. The excised mass from the knee measured 5×3 cm (Figure 5). Cut section showed brownish yellow shiny areas with intermixed white areas. It was solid and uniform. A small area of focal hemorrhage was seen but no necrotic areas were seen.



Fig 5: Specimen of the tumour after excision.

Histological section showed admixture of blood vessels, smooth muscle, and mature adipose tissue of varying proportions and distributions. Adipose tissue lobules consisted of fat cells with some variations in the cellular size and the perivascular mature smooth muscle cells were prominent. The blood vessels were thick-walled intermixed with longitudinal and transverse smooth muscle bundle (Figure 6). A diagnosis of Angiomyolipoma was made.

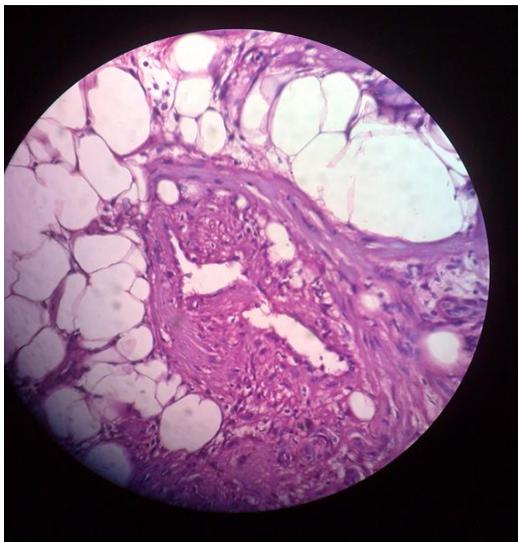


Fig 6: Histological section shows the tumour was composed of thick walled blood vessels, bundles of mature smooth muscle cells with a prominent perivascular arrangement and mature adipose tissue

Discussion

Angiomyolipoma is a rare complex mesenchymal neoplasm typically occur in the kidney and composed of fat, vascular, and smooth muscle elements. The amount of each component is variable. Most of the AMLs are benign, however rare aggressive variants are also reported. It has an incidence of about 0.3-3% [7]. There are two types of angiomyolipomas. One is isolated variant and another is associated with tuberous sclerosis. Eighty percent of renal AML occur sporadically and are often solitary [7].

They are common in females. AML belongs to PEComa

tumour family because they demonstrate perivascular epithelioid cell (PEC) differentiation [8]. PEComa tumor family also includes lymphangio myomatosis, clear cell tumor of the lung, and a group of rare, morphologically, and immunohistochemically similar lesions seen at other sites. Only 20% of renal AML occur in association with TS and usually multicentric [9, 10].

Extrarenal sites of angiomyolipomas are rare and occur most commonly in the liver; however, retroperitoneal soft tissue is the second most common location [1]. Angiomyolipoma in the musculoskeletal tissue particularly in the subcutaneous region is very rare. Clinically, these tumours are acquired, solitary and present as an asymptomatic nodule. These lesions are either subcutaneous or muscular and commonly found in extremities [3, 4, 5].

Hatori *et al.* reported a case of AML in the subcutaneous region of the popliteal fossa of the knee. Our case was also located in the subcutaneous tissue but at the level of the knee joint line in the medial aspect. To the best of our knowledge, this is the second case of a solitary angiomyolipoma in the subcutaneous tissue of the knee joint area without tuberous sclerosis or renal angiomyolipoma. Another case of multicentric intraarticular angiomyolipoma of knee joint and soft tissue of the foot was reported [10].

CT and MRI for angiomyolipoma is not diagnostic. However, Features of AML in MRI is quite peculiar with hypointense or hyperintense signals on T1 weighted images and heterogenous hyperintensity on T2 weighted images due to fat content [1, 10]. In our case MRI revealed an encapsulated, well defined fatty mass in the subcutaneous tissue with intermixed soft tissue densities. As the tumour is composed of adipose tissue, blood vessels and smooth muscle, MRI clearly demonstrated a tumor with mixed signal intensities. It was well enhanced on T1-weighted images and high signal intensities was seen on T2-weighted images.

Unusual tumors are often difficult to diagnose with standard clinical examination and radiological investigation and frequently a differential diagnosis is made. After histopathological examination from the biopsy of excised tumour final diagnosis is usually possible. For the diagnosis of angiomyolipoma, an admixture of small to medium-sized thick-walled blood vessels, smooth muscle cells and adipose tissue is required on histopathological section [9]. Similar features were seen in our case also. In some tumours adipose tissue is the predominant component, and in others smooth muscle predominated.

Histologically, AMLs should be differentiated from angioliipoma, angioleiomyoma and hemangioma with partial involution. In AMLs elastic tissue stains may reveal blood vessels which have developed an elastic lamina whereas other blood vessels lack it [9]. These unique features of this lesion distinguish it from other lesions. In angioliipoma and hemangioma, smooth muscle bundles are characteristically absent while angioleiomyoma is devoid of adipose tissue component.

There are different opinions regarding the origin of the cells of AMLs. Barnard *et al.* examined the cell origin of angiomyolipoma by electron microscopy and immunohistochemistry and stated that the AML is probably originated from a single cell that shares homology with the pericyte [2]. Immature short spindle and epithelioid cells in angiomyolipoma might be primitive mesenchymal cells with a capability to differentiate toward both smooth muscle and fat cells. Smooth muscle component of the tumour may be hypercellular, atypical or epithelioid and derived from the

vessel wall. Mature adipose tissue is seen in most of the tumours^[11].

The treatment of AML is wide local resection of the entire lesion, which is usually curative^[1, 10]. We excised the whole tumour en masse from the surrounding tissue. There was no adhesion or infiltration. Particularly important is the identification of any features of malignancy that changes management and prognosis markedly. Biopsy revealed no features of aggressive lesion in our case. There was no recurrence at 2 years and 5 years of follow-up at the operative site. Despite the uncharacteristic features, nearly all angiomyolipomas are benign and no recurrence usually occur. Malignant transformation, if it ever occurs, is extremely rare^[2, 10].

Conclusion

Extrarenal angiomyolipomas are complex mesenchymal tumours. These are usually solitary benign lesions and malignant transformation is exceedingly rare. AMLs in the subcutaneous tissue may mimic many other lesions. Diagnosis depends on clinical examination, imaging and histological examination. Wide resection of the tumour, which is usually curative. Our patient with AML in an unusual site of the knee, has had an uneventful post-operative course without any features of recurrence.

References

1. Ren N, Qin LX, Tang ZY *et al.* Diagnosis and treatment of hepatic angiomyolipoma in 26 cases. *World J Gastroenterol.* 2003; 9:1856-1858.
2. Hatori M, Watanabe M, Kokubun S. Angiomyolipoma in the knee—Acasereport. *Ups J Med Sci.* 2005; 110:245-9.
3. Krolick WD, Black JR. Angiomyolipoma in the foot. A case report. *J Am Podiatr Med Assoc.* 1987; 77:290-1.
4. Kuroda S, Itoh H, Yamagami T *et al.* Angiomyolipoma arising in the thigh. *Skeletal Radiol.* 2000; 29:293-7.
5. Pikoulis E, Bramis C, Michail O *et al.* Angiomyolipoma arising in the gluteal region. *Chin Med J (Engl).* 2007; 120:1284-6.
6. Insabato L, Rosa G, De Terracciano LM *et al.* Primary monotypic epithelioid angiomyolipoma of bone. *Histopathology.* 2002; 40:286-90.
7. Rakowski SK, Winterkorn EB, Paul E, Steele DJ, Halpern EF, Thiele EA. Renal manifestations of tuberous sclerosis complex: Incidence, prognosis, and predictive factors. *Kidney Int.* 2006; 70(10):1777-82.
8. Khan MS, Iram S, O'Brien TS *et al.* Renal 'perivascular epithelioid cellomas.' *BJU Int.* 2006; 98:1146-1147.
9. Hornick JL, Fletcher PE. Coma: what do we know so far? *Histopathology.* 2006; 48:75-82.
10. Varshney MK, Jain M, Sud A *et al.* Unusual multicentric angiomyolipoma of knee joint and soft tissue foot. *Joint Bone Spine.* 2011; 78:85-87.
11. Okada K, Yokoyama S, Nakayama I, Tada I, Kobayashi M. An electron microscopic study of hepatic angiomyolipoma. *Acta Pathol Jpn.* 1989; 39:743-749