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Angiomyolipoma in the Knee-A Case Report

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ABSTRACT

Extrarenal angiomyolipoma is an uncommon disease and this may be the first report of angiomyolipoma in the knee, mimicking a malignant sarcoma. A 38-year-old man without tuberous sclerosis presented with a history of increased mass in the knee joint. Computerized tomography and magnetic resonance imaging demonstrated a ÇS x ÇS x ÇR cm subcutaneous tumor in the knee joint. The tumor was widely excised. Histologically, the tumor was, well circumscribed, and composed of smooth muscle, vascular spaces, connective tissue, and mature fat. There were no signs of recurrence at one year and eight months after surgery.

INTRODUCTION

During the 1960's I made a series of inventions which ultimately led to the developAngiomyolipoma is a hamartomaous lesion, usually occurring either within or intimately associated with the kidney; pedunculated or satellite nodules may be found in the adjacent soft tissue and less commonly, in regional lymph nodes [1]. However, it is possible to find it in extrarenal sites, being the liver the most frequent one. The rest of sites recorded in literature are exceptional [2]. We describe a case of angiomyolipoma arising in the subcutaneous region in the knee with the radiological imaging and microscopic features.

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The patient was a 38 years old man. Five years ago, he noticed a small mass in his left popliteal fossa. The mass gradually increased in size. He visited a clinic and underwent surgery. The surgery became limited to incisional biopsy because of bleeding of the tumor. He was referred to us with the suspicion of sarcoma. At the examination, a mass was located in the popliteal fossa of the left knee. It had mild tenderness. No adhesion to the surrounding tissues was found. Computed tomography(CT) and magnetic resonance imaging(MRI) demonstrated a 4 x 4 x 3 cm sized mass in the subcutaneous region in the medial side of the popliteal fossa (Fig 1). The mass was widely excised. At one year and eight months after surgery, there were no signs of recurrence.

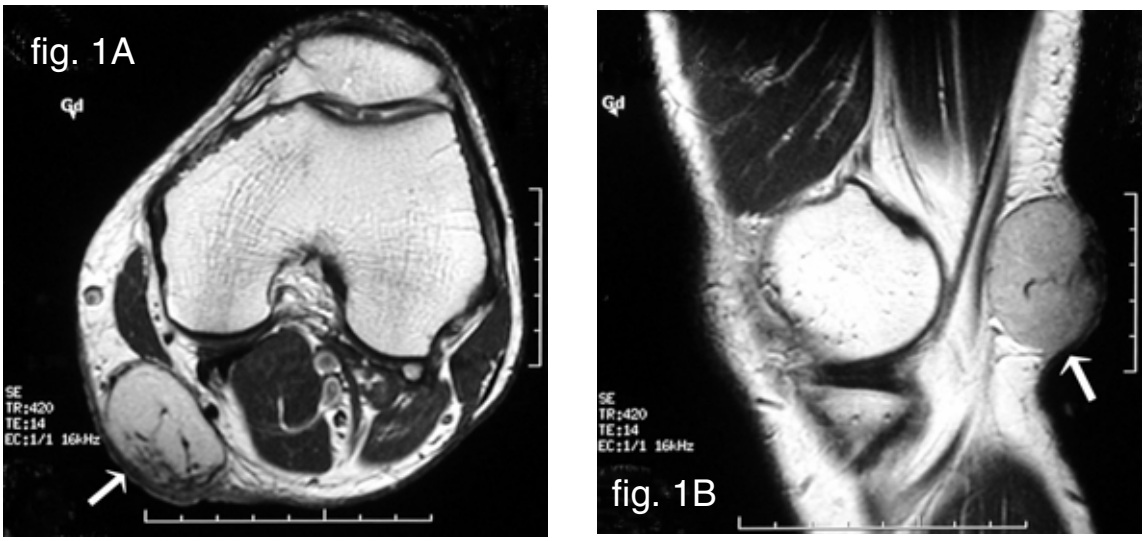

Fig. 1 MR images demonstrating a subcutaneous mass (arrow) which was well enhanced after gadolinium injection on T1 weighted images (A: axial image, B: sagittal image)

Grossly, the tumor presented as a whitish to slightly brown colored well-circumscribed mass in the subcutaneous region. Focal hemorrhage was present. Microscopically, the tumor was composed of convolutes of thick walled blood vessels, interlacing bundles of mature smooth muscle cells with a prominent perivascular arrangement and mature adipose tissue showing some variations in the cellular size. These three components were arranged irregularly and intermingled each other. Rich vascular channels of various sizes were present, some of which were dilated showing hemangioma-like appearances (Fig 2). Elastica Masson Goldner stain clearly showed prominent collagen fibers and focally elastic fibers surrounding smooth muscle cells. Immunohistochemically, the spindle cells in the tumor were reactive with HHF-35 and Éø smooth muscle actin. These positive spindle cells were found around the adipose tissue. The fat cells were positive for S100 protein,

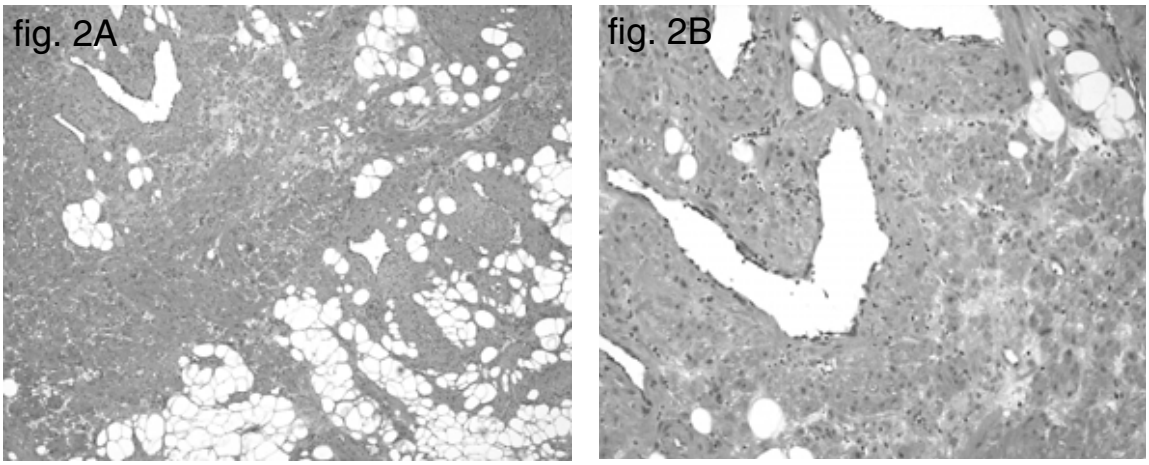

Fig. 2 Microscopically the tumor was composed of convolutes of thick walled blood vessels, interlacing bundles of mature smooth muscle cells with a prominent perivascular arrangement and mature adipose tissue showing some variations in the cellular size (A : low power view x1 , B : high power view x 10).

and the vascular endothelial cells were reactive with CD 34. No apparent reactivity was found for HMB-45.

DISCUSSION

Angiomyolipoma occurs most commonly in the kidney. The tumor is more common in women than in men. Angiomyolipoma is more often encountered in patients with tuberous sclerosis. Approximately one third to one half of cases are associated with tuberous sclerosis; less rarely with lymphangioma and lymphangiomatosis [1]. Extrarenal sites of angiomyolipoma include the liver [3, 4], spleen [5], retroperitoneum [6], lymph nodes [3] and spermatic cord [7]. The subcutaneous angiomyolipoma is very rare [8, 9,10,11]. In 1990, Fitzpatrick et al. described eight cases of cutaneous angiolipoleiomyoma (“angiomyolipoma”). Clinically, the tumors were acquired, solitary, asymptomatic nodules that were always acral in location. Patients' ages ranged from 33 to 77 years (median 52.6 years); the male/female ratio was 7:1. Signs of tuberous sclerosis or renal angiomyolipoma were absent in all these cases [9]. To our knowledge, this is the first case reported of an angiomyolipoma in the knee joint area in which tuberous sclerosis or renal angiomyolipoma was not seen.

Diagnostically, CT scan and MRI reveal a fatty mass with intermixed soft tissue densities, except in those cases in which absence of fat or hemorrhage obscures the radiological findings. The tumour is composed of fat, blood vessels and smooth muscle. The fat has a characteristic tissue density that often permits easy identification of an angiomyolipoma on CT [12]. Already in 1988, Uhlenbrock et al. reported that at MR imaging, most angiomyolipomas could be clearly characterized because of the depiction of intratumoral fat especially by the use of fat-suppression techniques [13]. In the present case, CT and MRI did not reveal fatty component of the

tumor. Recently, Ren et al. reported that MRI showed hypointensity or hyperintensity on T1-weighted images and heterogeneous hyperintensity on T2-weighted images [14]. In our case, MRI clearly demonstrated a tumor in the subcutaneous region with low signal intensities and well enhanced on T1-weighted images and high signal intensities on T2-weighted images.

Microscopically, the tumor is composed of three different tissue components that vary greatly in distribution: (1)mature adipose tissue, (2) convolutes of thick walled blood vessels, (3) irregular arranged sheets and interlacing bundles of smooth muscle [16]. In some tumors the fat is the predominant component, and in others smooth muscle predominated. Using Elastic tissue stains it is possible to reveal that some blood vessels have developed an elastic lamina whereas other blood vessels lack it [9]. These unique features of this lesion distinguish it from other lesions such as angiomyoma, angiolipoma, and other mixed mesenchymal tumors [15]. Angiomyolipomas show consistent immunopositivity for HMB-45 [16]. HMB-45 reactivity, reported for renal angiomyolipomas, has been suggested as a useful tool in differential diagnosis. Buyukbabani et al. conclude that, unlike renal angiomyolipomas, HMB-45 reactivity is not helpful in differentiating cutaneous angiomyolipomas [8].

The cell of origin remains mysterious. Barnard et al. analyzed angiomyolipoma by electron microscopy and immunohistochemistry to determine the appearance and nature of cells composing angiomyolipomas. The study demonstrated that the angiomyolipoma is likely derived from a single cell that shares homology with the pericyte [16]. Okada et al considered that the immature short spindle and epithelioid cells in angiomyolipoma might be primitive mesenchymal cells having an ability to differentiate toward both smooth muscle and fat cells [4].

Despite the atypical features, nearly all angiomyolipomas seem to pursue a benign clinical course. There is no evidence that the presence of regional or systemic lymph node involvement, perirenal satellite tumors, or angiomyolipomas growth in other organs reflects malignant potential. Malignant transformation, if it ever occurs, must be exceedingly rare [17, 18]. The present case, initially suspected as a sarcoma by unusual microscopic appearances in an unusual site of the knee, has had an uneventful course after surgery.

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