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Angiolipoma-like Atypical Lipomatous Tumor/Well-differentiated Liposarcoma

Anjiyolipoma Benzeyen Atipik Lipomatöz Tümör/İyi Diferansiye Liposarkom

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ABSTRACT

Lipomas are benign mesenchymal neoplasms. Angiolipoma is a variant of ordinary lipomas, characterized by adipocytic cells as well as capillary vascular clusters. Atypical lipomatous tumors are local aggressive mesenchymal neoplasms which include atypical stromal cells. They frequently arise in the deep thigh of the lower extremity. They have four histologic subtypes. So far, there aren't any case of angiolipoma with malignant transformation or atypical lipomatous tumor with angiolipoma-like areas in the English literature. Here we presented a case of atypical lipomatous tumor which was quite like angiolipoma. It was composed of mature fat with focal cytologic atypia and also accompanying diffuse capillary vessels. If there is a large-diameter and deeply located angiolipoma-like lesion, careful examination is required to rule out a liposarcoma. Liposarcomas may be rarely dedifferentiate or metastatic. It is not possible to verify that if the case is a variant of liposarcoma or it is a malignant transformation of angiolipoma.

Keywords: Atypical lipomatous tumor, liposarcoma, angiolipoma, malign transformation, angioliposarcoma

ÖZ

Lipomlar iyi huylu mezenkimal neoplazmlardır. Anjiyolipom ise adipositik hücrelerin yanı sıra kılcal damar kümeleri ile karakterize edilen sıradan bir lipom varyantıdır. Atipik lipomatoz tümörler atipik stromal hücreler ile karekterize lokal agresif mezenkimal neoplazmlardır. Sıklıkla alt ekstremitelerde derin yerleşimli olarak ortaya çıkarlar. Dört histolojik alt tipi mevcuttur. Şimdiye kadar, İngiliz literatüründe malign transformasyonlu herhangi bir anjiyolipom veya anjiyolipom benzeri alanlara sahip atipik lipomatöz tümör tanımlanmamıştır. Biz burada fokal sitolojik atipisi olan, matür yağ hücreleri ve eşlik eden yaygın kılcal damarlardan oluşan anjiyolipom benzeri bir atipik lipomatöz tümör olgusunu sunduk. Büyük çaplı ve derin yerleşimli anjiyolipom benzeri lezyonlarda, liposarkomu ekarte etmek için dikkatli inceleme yapmak gerekir. Liposarkomlar nadiren dediferansiye veya metastatik olabilir. Bizim olgunun liposarkomun bir varyantı mı yoksa anjiyolipomun malign transformasyonu mu olduğu konusu tartışmalıdır.

Anahtar kelimeler: Atipik lipomatöz tümör, liposarkom, anjiyolipom, malign transformasyon, anjiyoliposarkom

INTRODUCTION

Atypical lipomatous tumors are locally aggressive mesenchymal tumors. They are composed of mature fats with variably sized adipocytes, bands of fibrotic stroma, and also have atypical stromal cells. Areas of fat necrosis are observed, especially at the peripheral localization of the large lesions. These tumors are frequently observed in the deep locations of the thigh, followed by the retroperitoneum, trunk, head and neck region, and spermatic cord. They are predominantly seen in the middle and older ages. Atypical lipomatous tumors have four histologic subtypes, including lipoma-like, sclerosing, inflammatory, and spindle cell histologic subtypes. The lipoma-like subtype is the most common.

Diffuse capillary vascular structures are unusual event in atypical lipomatous tumor. This pattern is very similar to angiolipoma. Angiolipomas are benign tumors composed of mature adipocytes and intermingled with small and thin-walled vessels with intraluminal fibrin thrombi. Angiolipomas constitute 6%-17% of all lipomas (1). They were first described by Bowen (2) in 1912. The nature of these lesions was first documented in 1960 by Howard and Helwig (3). In 1974, Lin and Lin (1) divided them

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into two; infiltrating and non-infiltrating groups based on their biological behavior. Infiltrating angiolipomas are characterized by a non-capsulated tumor extended into surrounding tissues. Non-infiltrating angiolipomas are encapsulated.

İn this study, we presented a case of atypical lipomatous tumor composed of mature fat with focal cytologic atypia and diffuse capillary vessels. It was similar to angiolipoma. To our knowledge, it is probably the first case in English literature.

CASE PRESENTATION

A 75-year-old male patient was admitted to an orthopedic clinic with complaints of a palpable mass in his back. The mobile mass lesion under the latissimus dorsi muscle was excised. No metastatic lesion was detected by computed tomography (CT) scans. Macroscopic examination revealed that the mass lesion was covered by a thin incomplete capsule.

The cut surface of the lesion had a dirty yellow color, with an appearance of mature fat (Figure 1). Most of the neoplastic adipocytic cells had similar sizes and shapes (Figure 2A-C). There were also scattered capillary vascular clusters within the lesion and capillary vascular structures were usually located in the periphery of the lesion. Endothelial cells of capillary structures were benign. The adipocytes size in different parts of the lesion varied from small to large compared to normal (Figure 2D and Figure 3A, B). In addition, there were atypical stromal cells and rare lipoblasts in fibrous septa (Figure 3A-C). Atypic stromal cells rarely showed pleomorphism and there were sparse mitotic activities (Figure 3D). Immunohistochemically, capillary structures expressed CD34 (Figure 4A); atypical stromal cells expressed murine double minute 2 (MDM2), cyclin-dependent kinase 4 (CDK4), and p16 (Figure 4B, C); and scattered atypical stromal cells expressed S100 protein (Figure 4D). Analysis of MDM2 by fluorescence in situ hybridization (FISH) showed clusters of red signals in the neoplastic cells. Red signal clusters in neoplastic stromal cells indicated the presence of amplification/overexpression of the MDM2 protein (Figure 5).

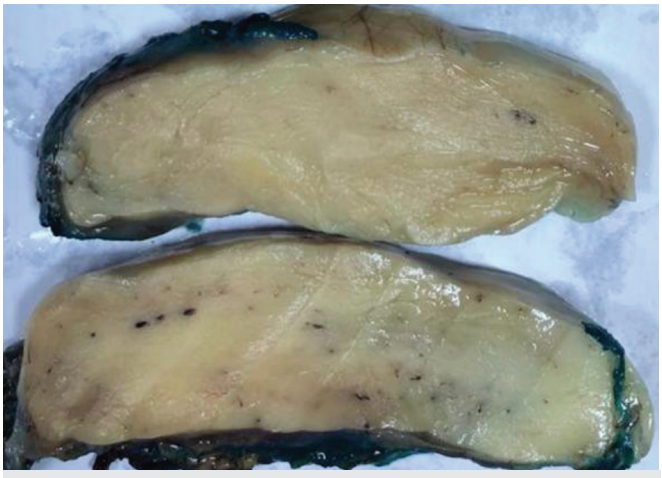

Figure 1. Tumor is covered by a thin fibrous capsule. The cut surface shows a dirty yellow color with a mature fatty tissue appearance

The patient was discharged without any complications and after ten months of follow-up, there was no recurrence as determined by endoscopic evaluation and CT examination.

Consent form was taken from the patient.

DISCUSSION

Angiolipoma is a variant of ordinary lipomas characterized by adipocytes as well as capillary vascular clusters. We did not find any case of angiolipoma with malignant transformation or an atypical lipomatous tumor with angiolipoma-like areas in the English literature. Due to the atypical stromal cells in the lipomatous areas and the presence of widespread capillary vessels, it may be misdiagnosed as angiolipoma. Differential diagnosis of angiolipoma depends on the density of the capillary vessels. Hypovascular angiolipomas are differentiated from

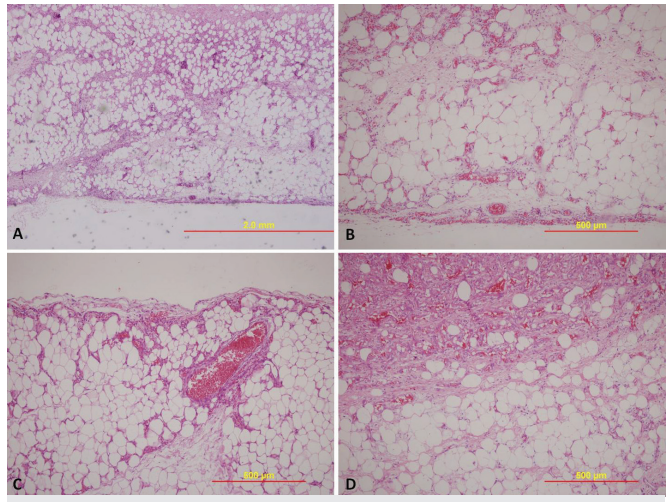

Figure 2. A, B, C) Hematoxylin and eosin (H&E), x100: Adipocytic cells are usually similar in size and shape, and there are some capillary vascular clusters scattered within the lesion. **D)** H&E, x200: Adipocytic cells have different sizes and shapes in some areas

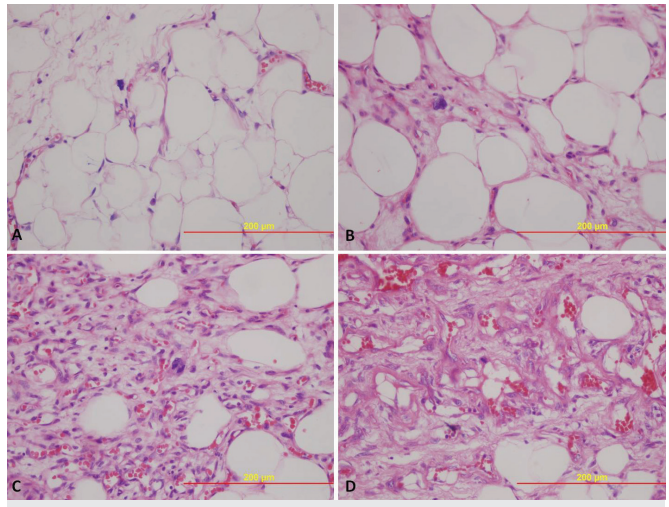

Figure 3. A, B, C) Hematoxylin & eosin (H&E), x400: Atypical stromal cells in the fibrous septa. **D)** H&E, x400: Capillary structures

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ordinary lipoma by the presence of fibrin thrombi, meanwhile cellular angiolipomas may be confused with Kaposi's sarcoma or spindle cell hemangioma. Although they do not contain clustered vessels with small thrombi, spindle cell hemangiomas unlike angiolipoma, contain large cavernous vascular structures and large calcified thrombi (phleboliths).

Infiltrative angiolipomas are localized in the deep soft tissue (1,4), not encapsulated, and can be mixed more often with angiosarcoma. Our case was located deeply under the muscle in the back, but it was circumscribed and had lobulated margins.

Angiosarcoma and Kaposi's sarcoma in this case were considered in the differential diagnosis due to the focal capillary vascular proliferation detected in the mature adipose tissue. There were fascicular endothelial proliferations consisting of more spindle cells than angiolipoma in Kaposi's sarcoma. There were also blood-filled slit-like spaces, extravasated erythrocytes, and periodic acid-Schiff [PAS (+)] hyaline globules in Kaposi's sarcoma, but no fibrin thrombi. Immunohistochemical studies showed that Kaposi's sarcoma endothelial cells expressed human herpesvirus 8. In angiosarcoma, the vascular structures dissected fatty tissue, collagen fibers, and other tissues. The endothelial cells of the vascular structures were atypical, multi-layered, and showed tufting toward the lumen. There were no fibrin thrombi in capillary vessels in angiosarcoma.

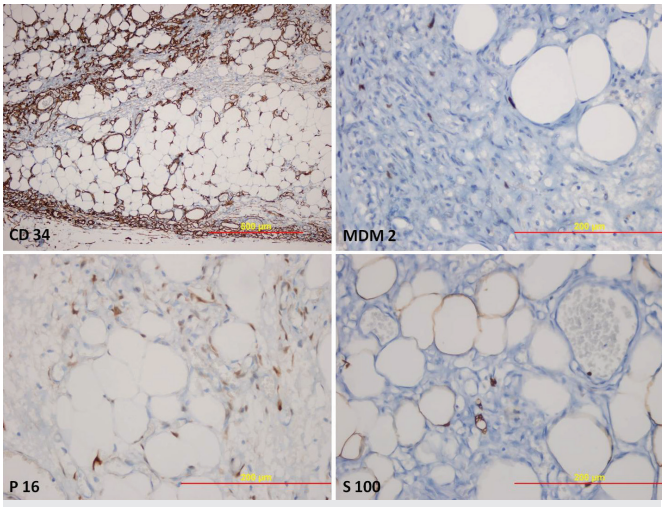

Figure 4. CD34x200: Capillary structures. MDM2, x400: Atypical stromal cells. CDK4, x400: Atypical stromal cells. P16: Atypical stromal cells. S-100x400: Atypical stromal cells
MDM2: Murine double minute 2, CDK4: cyclin-dependent kinase 4

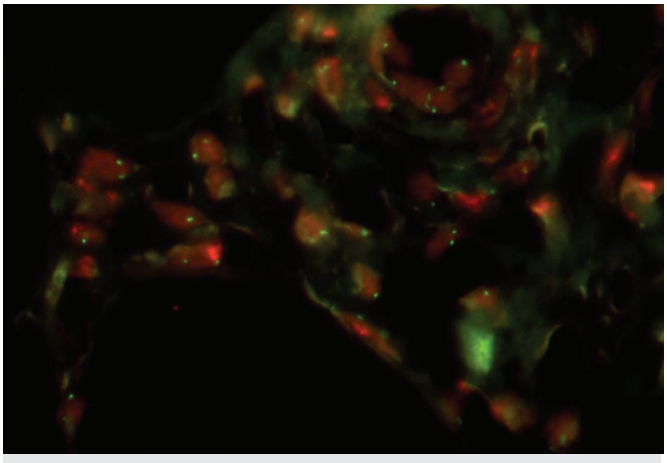

Figure 5. FISH: MDM2 amplification and overexpression
FISH: Fluorescence in situ hybridization, MDM2: murine double minute 2

Our case was diagnosed as angiolipoma-like atypical lipomatous tumor/well-differentiated liposarcoma due to the presence of diffuse capillary proliferations in the atypical lipomatous tumor.

The presence of atypical stromal cells is important to differentiate atypical lipomatous tumor from a lipoma. Thway et al. (5) had reported that p16, CDK4, and MDM2 were useful immunohistochemical markers for the distinction of lipoma and an atypical lipomatous tumor. In addition, we have also showed MDM2 amplification by FISH. MDM2 amplification is a finding that supports malignant transformation. The co-occurrence of angiolipoma and atypical lipomatous tumor has been reported by Christodoulidou et al. (6) in a case presentation. However, angiolipoma and atypical lipomatous tumors were seen in the paratesticular area and in contralateral localizations. However, in our case, angiolipomatous areas and atypical lipomatous tumor were intermingled with each other.

Fibrin thrombi are usually seen in angiomatous lesions, caused by microtrauma. They are usually seen in subcutanous angiolipomas and not in angiolipoma-like areas in our case. In the literature like in our case, angiolipomas in the gastric, colonic and rectal locations without fibrin thrombi have been reported (7-9). The absence of fibrin thrombi in the deep soft tissue may be associated with less trauma in these areas.

In our case, it was noticed that the adipocyte cells in fatty tissues, which contained dense capillary vessels showed different sizes and shapes. The diagnosis was atypical lipomatous tumor because of the presence of rare atypical stromal cells and lipoblasts in fibrous septa in these areas. Is the present case a new variant of liposarcoma? It can be discussed because of the diffuse capillary vessels similar to angiolipoma. However, some liposarcoma types have areas with branched capillaries, and a form with extensive capillary vascular proliferation has not yet been defined.

Can the atypical stromal cells and lipoblasts detected in the lipomatous areas in this case be the evidence of the malignant transformation of an angiolipoma? However, soft tissue textbooks report that angiolipomas are always benign or there is no evidence that these lesions ever undergo malignant transformation (10,11).

Angiolipoma-like atypical lipomatous tumors and angioliposarcoma or malignant transformation of angiolipoma have not yet been described in the literature. We defined our case as an atypical lipomatous tumor/well-differentiated liposarcoma due to the atypical stromal cells and lipoblasts observed in the angiolipoma-like and lipomatous areas. In conclusion, the possibility of atypical

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lipomatous tumor/well-differentiated liposarcoma in large and deeply located angiolipoma-like lesions should be excluded.

Informed Consent: Consent form was taken from the patient.

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