***Case Report* / Olgu Sunumu**

# A Rare Case of Angiomyxolipoma: Differential Diagnosis From Other Vascular and Myxoid Tumors

*Nadir Bir Anjiyomiksolipoma Olgusu: Diğer Vasküler ve Miksod Tümörler ile Ayırıcı Tanı*

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A case of angiomyxolipoma in the posterior neck region of a 36-year-old man is presented. Although lipomatous tumors are very frequent among benign soft tissue neoplasms, angiomyxolipoma has been reported only in seven case studies before. We present a new case of a typical angomyxolipoma. Histopathologically, admixture of mature adipocytes, poorly cellular myxoid spindle cell areas and abundant vascular structures are the striking components of this tumor. Myxoid spindle cell lipoma, vascular spindle cell lipoma, myxoid liposarcoma, myxolipoma, angiolipoma and pseudoangiomatous spindle cell lipoma can be considered in the differential diagnosis of angiomyxolipoma. Immunohistochemistry is usually helpful in the diagnosis of this extremely rare entity. In our case, positive staining for vimentin and sparse positivity for CD34, in the absence of reactivity for SMA, desmin, S-100 protein and HMB45 in the spindle cells, are the most important immunohistochemical features that help in the differential diagnosis.

*Key words:* Angiomyxolipoma; vascular; myxoid; tumor; immunohistochemistry; differential diagnosis.

Otuz altı yaşındaki bir erkek hastanın ensesinde ortaya çıkan bir anjiyomiksolipoma olgusu sunulmaktadır. Lipomatöz tümörlere benign yumuşak doku neoplazileri arasında oldukça sık rastlansa da, anjiyomiksolipom tanısı ile sadece yedi olgu sunumu vardır. Burada tipik anjiyomiksolipom olan yeni bir olguyu sunmaktayız. Histopatolojik olarak bu tümörün en çarpıcı özelliği birbiri içine giren matür yağ dokusu ve hücreden fakir miksoid iğsi hücreli alanlar ve eşlik eden çok sayıda damar yapısıdır. Miksoid iğsi hücreli lipom, vasküler iğsi hücreli lipom, miksoid liposarkom, miksolipom, anjiyolipom ve psödoanjiyomatöz iğsi hücreli lipom anjiyomiksolipom ayırıcı tanıya girebilecek olan lezyonlardır. Bu son derece nadir lezyonun ayırıcı tanısında immunohistokimya oldukça destekleyicidir. Bizim olgumuzda iğsi hücrelerdeki SMA, desmin, S-100 protein ve HMB45 negatifliği ve bunun beraberinde vimentin pozitifliği ve CD34 zayıf pozitifliği tanıya yardımcı olan immunohistokimyasal bulgular olmuştur.

*Anahtar sözcükler:* Anjiyomiksolipom; vasküler; miksoid; tümör; immunohistokimya; ayırıcı tanı.

Lipoma is a benign tumor composed of mature white adipocytes and is the most common soft tissue mesenchymal neoplasm in adults.[1] There are several forms of lipomas. Ordinary lipomas constitute approximately 80% of all lipomas, while other types such as angiolipoma, angiomyolipoma, spindle cell/pleomorphic lipoma, myolipoma, intramuscular lipoma make up the remaining 20% of lipomas.[2,3]Angiomyxolipoma was not identified as a distinct type of lipoma until Mai et al.[4] reported the first case of angiomyxolipoma (vascu-



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lar myxolipoma) in 1996. Since then, only seven other angiomyxolipomas have been reported.[5-9] We report here the eighth case of angiomyxolipoma and discuss the differential diagnosis.

## CASE REPORT

A 36-year-old man with a slowly growing painless mass located on the posterior neck, developing in a two-year period, is presented. Clinical examination revealed a 5x4 cm, solitary, slightly mobile subcutaneous nodule with a somewhat firmer consistency than that of a lipoma. Surgical excision of the mass was rather problematic as it was mostly composed of a mucoid material. Over the following six months after the surgery, no recurrence was documented.

Grossly total dimensions of four pieces of the yellowish-white, soft, shapeless, vascular, semi-solid gelatinous mass was measured as 62x52x14 mm (Fig. 1). Microscopically, the tumor consisted of an admixture of highly vascular and poorly cellular myxoid and lipomatous areas (Fig. 2). The myxoid areas occupied nearly 70% of the total tumor volume. These areas contained cells with oval or spindle shaped nuclei without atypia, nucleolus or mitosis. The dendritic cytoplasmic processes within the myxoid ground substance were highlighted by vimentin immunohistochemically. Alcian blue (pH 2.5) and mucicarmine stains showed diffuse strong staining in the myxoid areas. Scattered mast cells and histiocytes were found in the myxoid ground substance.

The lipomatous component of the tumor was randomly scattered throughout the tumor and was composed of mature fat cells without any atypia. No lipoblasts were found.

There were abundant thick-walled and fewer thin-walled blood vessels. Hyalinization of some of the thick-walled vessels was highlighted with Masson's trichrome stain. This stain also revealed scarce thin collagen fibers in the myxoid areas. Immunohistochemistry showed diffuse positivity for vimentin and scattered sparse positivity for CD34 in the cytoplasm of the spindle cells. HMB45, S-100 protein, alpha smooth muscle actin (SMA) and desmin were all negative in the spindle cells. Smooth muscle actin stained some of the walls of the thick-walled vessels while all vessels showed diffuse positivity for CD34, factor VIII and CD31. Mature adipocytes stained strongly for S-100 protein.

***Fig. 1.*** *Gross appearance of shapeless mucoid mass with punctuated vascular structures.*

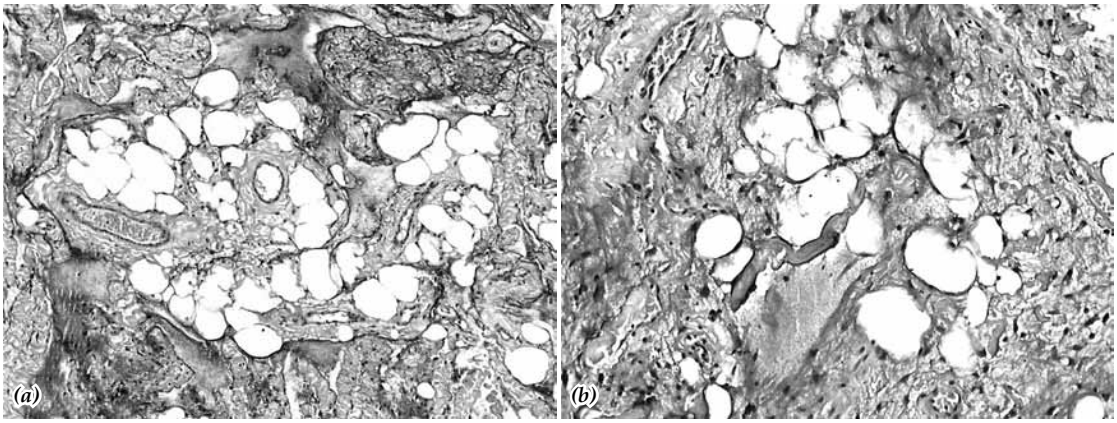


***Fig. 2.*** *Lipomatous lesion with abundant myxoid areas and numerous thin- and thick-walled vessels* ***(a)*** *(HE x 50),* ***(b)*** *(Mucicarmine x 50).*

## DISCUSSION

Although lipomatous tumors are very frequent among benign soft tissue neoplasms, angiomyxolipoma has been reported only in seven case studies before, thus the presented case is known to be the eighth one. Of these seven cases, two were in the scalp, two were in the extremities, one was in the thigh, one was in the spermatic cord and one was in the subungual region.[4-9] Our case is the thirrd one which is seen in the head and neck region. Except for one, all cases including ours were male. Ages of the previous cases ranged from 32 to 66 years.

Besides its characteristic histologic features, differential diagnosis of angiomyxolipoma is wide and to some extent it can be challenging. Histopathologically, the tumor is composed of an admixture of mature adipocytes, poorly cellular myxoid spindle cell areas and abundant vascular structures. There are both thin- and thick-walled vessels, some of which may show prominent hyalinization.[7,8]

The possibility of entrapment of the adjacent adipose tissue by an angiomyxoma or by a reactive myxoid vascular lesion is ruled out by random location of the scattered adipose tissue throughout the lesion.[7] The negativity for S-100 protein immunohistochemically also proved that, there were no entrapped nerve fibers. The characteristic histomorphology, distribution and abundance of the blood vessels also lead us to exclude preexisting vascular structures.[7]

Immunohistochemistry and histopathology alone can be considered as sufficient methods for the correct diagnosis. However, myxoid spindle cell lipoma is especially important in the differential diagnosis. A previously reported vascular variant of spindle cell lipoma (SCL) makes the situation more challenging.[7] Chromosomal analysis of the spindle cells in most of the SCLs shows an unbalanced aberration involving 16q resulting in monosomy or partial loss of 16q, but others have demonstrated abnormalities of 13q and 6p.[3] On the other hand, a cytogenetic analysis of an angiomyxolipoma revealed translocations t(7;13)(p15;q14) and t(8-12)(q12;p13). Those results showed engagement with chromosomal regions involved in certain benign adipose and myxoid tumors. Thus, it can be considered that angiomyxolipoma may share related chromosome aberrations with SCL.[6]

In our case, positive staining for vimentin and sparse positivity for CD34, in the absence of reactivity for SMA, desmin, S-100 protein and HMB45 in the spindle cells, are the most important immunohistochemical features that help in the differential diagnosis. In case of 'diffuse' positivity for CD34 with fewer amounts of vascular structures and presence of extensive 'ropy' collagen bundles, the diagnosis can likely be 'myxoid SCL'.[2,7]

In the ultrastructural study of the first case of angiomyxolipoma, reported by Mai et al.,[4] the satellite cells showed long cellular processes in the background of loose electron dense material, transversed by scattered bundles of collagen. Thus it is very difficult to build up the differential diagnosis mainly on the absence of collagen fibers. In the presented case, though it is not ropy, Masson's trichrome stain showed some sparsely scattered thin collagen fibers in between the spindle cells of the myxoid areas.

Vascular SCL is differentiated from angiomyxolipoma by the absence of myxoid areas and presence of other features of SCL.[8] Pseudoangiomatous SCL lacks both real vascular structures and myxoid stroma.[3,8] Myxolipoma, another entity in the differential diagnosis of angiomyxolipoma, does not have a prominent vascular component; similarly angiolipoma and angiomyolipoma do not have myxoid ground substance.[3,8]

Myxoid liposarcoma should also be kept in mind in the differential diagnosis of angiomyxolipoma. However, it is not a problematic lesion as it does not have the abundant thin- and thick-walled vascular structures of angiomyxolipoma. Rather, it has the characteristic chicken-wire-like plexiform capillary network with variable numbers of small lipoblasts.[1,3]

As a conclusion, angiomyxolipoma is a very rare, distinct benign lipomatous tumor which should be distinguished from other lipomatous or myxoid tumors. Its characteristic histopathologic features and immunohistochemical profile are of great importance for the differential diagnosis of this rare entity.

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