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Caso aislado

Angiomyxolipoma: A variant of lipoma

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RESUMEN

Presentamos un caso de angiomixolipoma en el tejido subcutáneo de la espalda superior en un varón de 50 años. En el examen clínico, la lesión se observa móvil y ligeramente dolorosa. La muestra patológica es redondeada, bien circunscrita, blanda y con una superficie pardo amarillenta y gelatinosa. Histológicamente la lesión consiste en tejido adiposo y estroma mixoide que contiene numerosos vasos sanguíneos de pequeño y mediano tamaño. Ésta es una variante poco frecuente de lipoma, que fue descrita por primera vez en 1996 en la zona de cuerda espermática. La consideramos un híbrido entre angiolipoma y lipoma de células fusiformes. La lesión debería distinguirse del liposarcoma mixoide por razones terapéuticas y pronósticas. **Rev Esp Patol 2000; 33(1): 41-45.**

Palabras clave: Angiomixolipoma - Lipoma - Tejido subcutáneo - Lipoma de células fusiformes

SUMMARY

We report a case of angiomyxolipoma in the subcutaneous tissue of the upper back of a 50-year-old man. Clinically, the lesion was mobile and slightly tender. The pathological specimen was rounded, well circumscribed, rubbery with a brownish-yellow gelatinous cut surface. Microscopically, the lesion consisted of adipose tissue and a myxoid stroma containing numerous small- and medium-sized blood vessels. This rare variant of lipoma was first described a year ago in the spermatic cord area. We consider it to be a hybrid between angiolipoma and spindle cell lipoma. The lesion should be distinguished from myxoid liposarcoma for obvious therapeutic and prognostic reasons. **Rev Esp Patol 2000; 33(1): 41-45.**

Key words: Angiomyxolipoma - Lipoma - Subcutaneous tissue - Spindle cell lipoma

INTRODUCTION

Several variants of lipoma have been described depending on the various mesenchymal components associated with the adipose tissue. Such components include myxoid, chondroid, vascular, smooth muscle and fibroblastic tissue (1). Several combinations of these have already been described. Herein we describe a case of angiomyxolipoma. This lesion, which was first described and named two years ago (2), has vascular, myxoid and lipomatous components.

CLINICAL HISTORY

A 50-year-old man presented with a 3-year history of a mobile, slightly tender mass in the upper back. Physical examination demonstrated a soft 3-cm subcutaneous nodule. Complete extirpation of the nodule was carried out.

MATERIALS AND METHODS

The specimen was routinely fixed in 10% buffered formalin and embedded in paraffin. Representative 4-µm sections were stained with hematoxylin and eosin, colloidal iron and periodic acid-Schiff stains. Immunohistochemical staining was performed on paraffin sections using a streptavidin-biotin-peroxidase technique. The following antibodies were applied: vimentin (Novocastra, clone V9, dilution 1:800), *Ulex europeus* (Vector Laboratories, dilution 1:1500), CD34 (Becton Dickonson, clone MY 10, dilution 1:200), S-100 protein (Biogenex, polyclonal, dilution 1:1600), muscle-specific actin (MSA) (Enzo Diagnostics, clone HHF35, dilution 1:600) and HMB-45 (Biogenex, clone HMB-45, dilution 1:400).

PATHOLOGIC FINDINGS

Gross examination of the specimen revealed a rounded, well-circumscribed, rubbery subcutaneous mass with a smooth external surface. The cut surface had a gelatinous appearance and a brownish-yellow color (Fig. 1). A well-defined capsule was not observed. Microscopically, the mass was completely surrounded by a thin layer of hypocellular fibrous tissue. Several medium-sized blood vessels transgressed this fibrous layer. The tumor consisted of a fairly even mixture of adipose tissue and a highly vascularized myxoid stroma (Fig. 2). The adipose tissue was slightly more concentrated towards the center of the lesion. The constituent adipocytes were fully mature and presented empty (lipid-filled) cytoplasm and pyknotic peripheral nuclei, which were indistinct at scanning magnification. No cell with lipoblastic features could be identified. The intercellular matrix had a basophilic granular appearance due to a high content of acid mucopolysaccharides. It stained positively with colloidal iron. The myxoid tissue consisted of several spindled and stellate cells with eosinophilic cytoplasm and ovoid or rounded vesicular nuclei (Fig. 3). Neither mitosis nor atypia was noted in these cells. Thin serpentine-shaped collagen bundles were also present but these were very focal and inconspicuous. Numerous small- and medium-sized blood vessels were seen evenly distributed throughout the lesion. The large majority of the vessels had walls at least 2- or 3-cells thick, with considerable quantities of collagen in-between the cells. The minority of the vessels consisted of capillaries; however, these were very few in

Figure 1. The tumor is well circumscribed, has a gelatinous appearance and is more yellowish toward the center.

Figure 2. Note the even mixture of the angiomyxoid and lipomatous components. Hematoxylin and eosin (original, ×12.5).

Figure 3. Higher magnification of Fig. 2 showing small blood vessels and the spindle cells of the myxoid tissue. Hematoxylin and eosin (original, ×300).

number and lacked the plexiform arrangement seen in myxoid liposarcoma. No evidence of tumor necrosis, hemorrhage, thrombosis or lymphocytic infiltration was observed. The overlying skin and adjacent subcutaneous tissue did not show any morphological alteration.

Positive immunoreactivity (Table 1) was observed with the following antibodies: vimentin in all the cells, *Ulex europeus* in the endothelial cells, CD34 in the endothelial cells as well as the spindle and stellate cells of the myxoid component (Fig. 4), S-100 protein in the adipocytes (Fig. 4), and muscle-specific actin (MSA) in the blood vessel wall. Immunostaining with antibody HMB-45 was negative.

DISCUSSION

This tumor showed a characteristic brownish-yellow gelatinous cut surface. The subcutaneous location in the

Table 1. Summary of the immunohistochemical results.

Antibody	Adipocytes	Vascular smooth muscle cells	Endothelial cells	Spindle and stellate cells
Vimentin	+	+	+	+
S-100	+	–	–	–
Muscle-specific actin	–	+	–	–
Ulex europeus	–	–	+	–
CD34	–	–	+	+
HMB-45	–	–	–	–

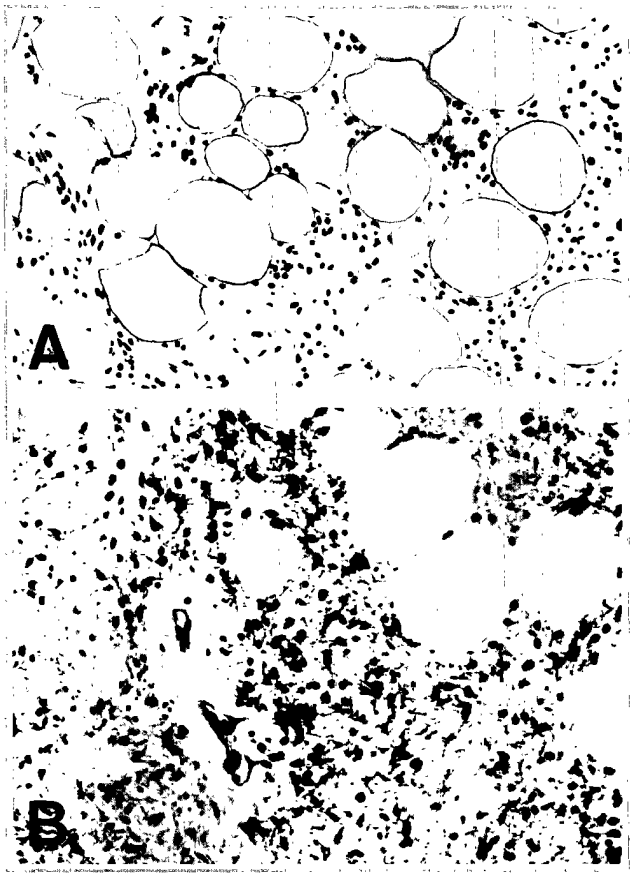

Figure 4. Mature adipocytes showing cytoplasmic and nuclear S-100 protein positivity (A), and the interstitial spindle and stellate cells and endothelial cells showing CD34 positivity (B) (original, ×100).

upper back, complete circumscription, presence of mature adipose tissue, and absence of lipoblasts all favor the diagnosis of a variant of lipoma.

Myxoid liposarcoma has many common features with the lesion described as angiomyxolipoma (AML) and is therefore the most important tumor to be ruled out for prognostic and therapeutic reasons. Similar to angiomyxolipoma, myxoid liposarcoma also has a myxoid stroma, lacks mitoses, may be well circumscribed by fibrous tissue and lacks significant cellular atypia. However, the data supporting a benign tumor in this case include the absence of lipoblasts, absence of a plexiform capillary pattern, the subcutaneous location and the age of the patient. Also, the myxoid stroma seen in this lesion had a granular basophilic appearance in contrast with the usually homogenous or glassy look of the stroma in myxoid liposarcoma.

Among the variants of lipoma, angiomyxolipoma combines features of angiolipoma (3) and spindle cell lipoma. Similar to angiomyxolipoma, spindle cell lipoma is composed of a mixture of mature fat cells and uniform spindle cells that are closely associated with myxoid matrix and a varying number of birefringent collagen fibers. However, angiomyxolipoma differs by containing in addition a very conspicuous vascular component consisting of small- and medium-sized vessels. We believe that the lesion reported by Warkel *et al.* (4) in 1982 and classified as an unusually vascular spindle cell lipoma may correspond to what we now call angiomyxolipoma. The first case of angiomyxolipoma was described in the spermatic cord area (2), a location quite unusual for the above mentioned variants of lipoma. Our case presented in the subcutaneous tissue of the trunk, a more typical region for these variants of lipoma. This therefore adds more evidence to the authenticity of angiomyxolipoma as a true and well-defined entity and variant of lipoma. Zamecnik (5) has recently described a case of angiomyxolipoma excised in the subcutaneous tissue of the temporal region of the scalp in a 57-year-old man that seems grossly and histologically identical to our case. No recurrence has so far been recorded in that case after 8 years of follow-up.

Angiomyolipoma, angiomyofibroblastoma (6), aggressive angiomyxoma (7, 8), cellular angiofibroma (9) and arteriovenous hemangioma can easily be ruled out if attention is paid to the three clearly defined components of this tumor (vascular, myxoid and lipomatous). Unlike our case, angiomyolipoma is HMB-45 immunopositive, usually has a renal location, and presents an extravascular smooth muscle component. Angiomyofibroblastoma, aggressive angiomyxoma, cellular angiofibroma (10), and arteriovenous hemangioma are not circumscribed and lack a significant lipomatous component.

The CD34 positivity observed raises a possible differential diagnosis with dermatofibrosarcoma protuberans, a borderline tumor that primarily affects the dermis and often invades the subcutaneous tissue (and for this reason, may be seen engulfing adipose tissue). However, unlike angiomyxolipoma, dermatofibrosarcoma protuberans has infiltrative borders, is rarely myxoid, contains radial whorls of spindle cells that produce a typical cartwheel pattern, and usually demonstrates a moderate to high mitotic activity. A previous work studied the CD34 positivity in a population of 90 lipomatous tumors (11) and their results showed that CD34-positive cells could be identified in all lipomas and their benign

variants. In this series, the spindle cell component of all cases of spindle cell lipoma was strongly positive for this antigen. Hence the CD34 positivity of the spindle cells in our lesion is not an entirely new or strange finding in lipomatous tumors and also constitutes further evidence that angiomyxolipoma is another variant of benign lipoma.

In conclusion, we present a lesion, which is identical to an previously described lesion called angiomyxolipoma (2), and thus favors this diagnosis. It is a highly vascular variant of spindle cell lipoma with a myxoid stroma. The location of this lesion in the subcutaneous tissue of the back and its CD34 positivity supply further evidence that angiomyxolipoma is truly a variant of lipoma. It must be distinguished from myxoid liposarcoma, which is a malignant tumor with possible recurrence and metastasis after excision.

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