CASE REPORT

Angiomyxolipoma; Report of a new case in subcutaneous tissue and review of the literature

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Abstract:
Angiomyxolipoma is considered as a variant of lipoma admixed with myxoid stroma and blood vessels. It was first described in 1996 by Mai and since then, 16 cases have been reported in literature, mostly in subcutaneous tissue. We report the case of a 50-year-old male who presented with a painless swelling of the right thigh. The ultrasonography revealed a vascularized inhomogeneous, slightly hyperechoic well defined mass measuring a maximum of 8 cm across. The patient underwent a large excision. The diagnosis was retained on histological and immunohistochemical findings. Angiomyxolipoma is an extremely rare benign tumor. This entity has to be differentiated from other benign and malignant lesions, the latter being much more common.

Key words: angiomyxolipoma, histopathology, subcutaneous tissue.

INTRODUCTION

Lipoma is a benign tumor composed of mature adipocytes. It is the most common soft tissue mesenchymal tumor in adults. It has many variants. Angiomyxolipoma (AML), or vascular myxoma, is considered as a variant of lipoma. It is a very rare benign tumor, first described in 1996 by Mai et al. (1). To date, 16 cases have been reported, located in subcutaneous tissue (2-9), spermatic cord (1), subungual area (10), intra-articular (11, 12), oral cavity (13), colon (14), and posterior mediastinum (15).

The aims of this article are to report a new case of angiomyxolipoma in the subcutaneous tissue, to discuss its differential diagnosis and to describe its follow-up.

CASE REPORT

We report a 50 year old man who presented with a two-year history of a painless swelling on the right thigh. Per examination, there was a relatively well-demarcated subcutaneous mass, firm and bosselated, which measured 10cm of large diameter. Sonography was performed, and revealed an inhomogeneous, slightly hyperechoic well defined mass lesion measuring a maximum of 8 cm across. This lesion is vascularized with arterial and venous flow.

The patient underwent a large excision of the mass. Grossly, the excised specimen measured 9 x 6.5 cm and was encapsulated. On cut surface, it was yellow-colored and appeared gelatinous in some areas. Histopathology demonstrated mature lipomatous tissue with areas of myxoid stroma containing numerous blood vessels of varying sizes (fig. 1). There is no atypia in myxoid stroma (fig. 2).

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Fig. 1: mature lipomatous tissue with areas of myxoid stroma containing numerous blood vessels (HE x 40).

Fig. 2: spindle cells in the myxoid component without atypia (HE x 100).

Immunohistochemical studies found that the spindle cells in the myxoid component expressed CD34, vimentine and smooth muscle actin (SMA). The mature adipocytes were positive for S100 protein and the blood vessels were immunoreactive for CD34 and SMA (fig. 3). Ki67 reaction was performed and was under 1%.

Fig. 3: spindle cells and blood vessels were immunoreactive for SMA (IHC x 40).

DISCUSSION

AML is a very rare benign tumor. It is a rare variant of lipoma with characteristic histopathologic and immunohistochemical features (14). It was first described by Mai et al. in 1996 (1), who reported the case of a 34 year-old man who has a tumor in the spermatic cord. This tumor was a proliferation of matures adipocytes associated with a myxoid stroma and with multiple blood vessels. After this, 15 cases have been reported in many sites. The patients’ ages ranged from 4 to 69 years. There is a male predominance with a sex ratio of 3 (table 1). The clinical presentation of AML is variable depending on the location. However, in soft tissue, it is most often a painless slowly growing mass (8, 11, 13).

Radiologically, the images are not specific (15). They can evoke a true lipoma, or more often a heterogeneous adipose tissue (11, 12, 15). Grossly, AML presents as a well-defined mass, of firm consistency. On cut section, it appears yellow-colored, lipomatous and gelatinous in some places, admixid with focal hemorrhage or dilated blood vessels (11-14). On microscopy, this tumor is composed of an admixture of myxoid areas with mature adipose tissue and numerous walled vessels. Immunohistochemical studies show that the cells in myxoid component express CD34 and vimentine. The mature adipose cells are immunoreactive for S100 protein whereas the blood vessels are positive for CD34, SMA and vimentine (15). However, Desmin and HMB45 were tested in many cases and were negative (table1). Ki67 reactions were tested by Lee et al. in 2 tumors. In both cases, Ki67 was under 3%.

Immunohistochemical studies in our case found, also, that Ki67 was less than 1%.

Differential diagnoses arise with both malignant and benign lesions. The main differential diagnosis is myxoid liposarcoma (14). This highly malignant lesion is much more common than AML. Therefore, it has to be diagnosed for prognostic and therapeutic reasons. In some difficult cases, myxoid liposarcoma is well circumscribed by fibrous tissue, mitoses are scanty and cellular atypia are mild. In these cases, the presence of lipoblasts and a plexiform or “chicken wire” vascular pattern are the key of diagnosis. If the tumor is hypervascular, the alternative possibility of low-grade myxofibrosarcoma should be considered (13). However, AML lacks the pleomorphism, nuclear atypia, the hypercellular and curvilinear vascular pattern of low-grade myxofibrosarcoma.

Other histological differential diagnosis includes benign lesions such as spindle cell lipoma, angiomyolipoma and angiolipoma (9). It is the
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Table 1: Clinical features and immunoprofile of reported cases of angiomyxolipoma

<table>
<thead>
<tr>
<th>Case No</th>
<th>Ref No</th>
<th>Sex/Age (years)</th>
<th>Location</th>
<th>Duration</th>
<th>CD34</th>
<th>Vimentine</th>
<th>SMA</th>
<th>Desmin</th>
<th>HMB45</th>
<th>K67</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1</td>
<td>M/53</td>
<td>Spermatic cord</td>
<td>3 months</td>
<td>NP</td>
<td>+/(s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>M/57</td>
<td>Scalp</td>
<td>NS</td>
<td>NP</td>
<td>+/(s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>M/50</td>
<td>Back</td>
<td>3 years</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>F/51</td>
<td>Thigh</td>
<td>4 months</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
<td>M/66</td>
<td>Scalp</td>
<td>NS</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
<td>-3%</td>
</tr>
<tr>
<td>6</td>
<td>6</td>
<td>M/44</td>
<td>Arm</td>
<td>7 years</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
<td>NP</td>
</tr>
<tr>
<td>7</td>
<td>7</td>
<td>M/57</td>
<td>Wrist</td>
<td>2 years</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
<td>NP</td>
</tr>
<tr>
<td>8</td>
<td>8</td>
<td>M/42</td>
<td>Subungual area</td>
<td>1 year</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>9</td>
<td>9</td>
<td>M/38</td>
<td>Gluted area</td>
<td>3 years</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>10</td>
<td>9</td>
<td>M/69</td>
<td>Hip</td>
<td>3 years</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>11</td>
<td>10</td>
<td>M/59</td>
<td>Intra articular</td>
<td>5 years</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>12</td>
<td>11</td>
<td>M/12</td>
<td>Buccal Mucosa</td>
<td>4 years</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>13</td>
<td>12</td>
<td>F/15</td>
<td>colon</td>
<td>===</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
<td>NP</td>
</tr>
<tr>
<td>14</td>
<td>13</td>
<td>F/53</td>
<td>Intra articular</td>
<td>Many years</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
<td>NP</td>
</tr>
<tr>
<td>15</td>
<td>14</td>
<td>M/41</td>
<td>Planter</td>
<td>NI</td>
<td>NI</td>
<td>NI</td>
<td>NI</td>
<td>NI</td>
<td>NI</td>
<td>NI</td>
</tr>
<tr>
<td>16</td>
<td>15</td>
<td>F/49</td>
<td>Posterior Medistium</td>
<td>Few weeks</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>17</td>
<td>Presen t case</td>
<td>M/30</td>
<td>Thigh</td>
<td>2 years</td>
<td>NP</td>
<td>+/(v,s)</td>
<td>+/(l)</td>
<td>+/(v)</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

SMA: Smooth muscle actin, F: Female, M: Male, S: Spindle cells in myxoid area, L: Lipocytes in lipomatous area, V: Vessel walls, NI: Not informed or not specified, NP: Not performed

The combination of the following three components: adipose tissue, blood vessels and the myxoid stroma favours the AML diagnosis. Chromosome analysis performed by Sciot et al. (4) in 2001 revealed that AML shares cytogenetic changes with lipoma, spindle cell lipoma and myxoma. The changes consist in translocations t (7;13)(p15;q13) and t (8;12)(q12;13)) (4, 8).

In summary, we present a new case of subcutaneous angiomyxolipoma of the thigh. Further cases are needed to more evaluate histopathologic features of these tumors. In our case, the treatment was surgical. Like all other cases, there was no recurrence.

REFERENCES

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