Case report

Aneurysmal (angiomatoid) fibrous histiocytoma

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We report a patient with aneurysmal (angiomatoid) fibrous histiocytoma manifested clinically as a single firm subcutaneous nodule, diagnosed as an epithelial cyst. Histologically, the growth showed massive histiocytic proliferation and areas with a vascular component. The lesion was histologically mistaken for Kaposi's sarcoma.

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Case report

A twenty-six-year-old white man was seen for a nodular lesion on the left side of his neck of 6 years duration. Initially, it was asymptomatic, but later it became tender and painful. The growth was subcutaneous, covered with normal skin, well defined, firm and rubbery in consistency, and measured 1.5 x 1.0 x 0.7 cm in diameter. All laboratory tests, including blood counts, blood chemistry, chest X-ray and liver function tests were within the normal range. VDRL was non-reactive. The lesion was surgically excised under local anesthesia.

Histopathology

Paraffin-embedded tissue sections were stained with hematoxylin and eosin, Masson's trichrome, Wilder's reticulum and the Prussian blue reaction for hemosiderin. The histologic sections showed a well-defined and solid nodular growth which appeared to be situated in part within a cystically dilated and thick-walled blood vessel. The muscular wall of the involved vessel was preserved in one half of the lesion. In this area, sections stained by the trichrome technique showed the muscle bundles very clearly within the vascular wall which was separated from the intravascular nodular growth by an area of cleft formation (Fig. 1). The solid intravascular growth was made up of massive proliferation of histiocytic cells (Figs. 2–3). Some of these cells showed abundant foamy cytoplasm. Others contained hemosiderin granules. A few histiocytic cells at the periphery of the growth showed large and hyperchromatic nuclei and exhibited a tendency for the formation of multinucleate giant cells (Fig. 4). There were no mitotic figures. Other cells present included scattered lymphocytes, plasma cells and eosinophils. At the periphery were a number of endothelial lined blood vessels with proliferation of elongated fibroblasts and foci of extravasation of erythrocytes (Fig. 5).
The growth is situated within an aneurysmal thick-walled blood vessel from which it is separated by an empty cleft (H&E, ×100).

In most parts the lesion is made up of a solid proliferation of histiocytic cells, some with foamy cytoplasms (H&E, ×225).
Fig. 3. Shows several foamy histiocytic cells and foci of extravasation of erythrocytes (H&E, x400).

Fig. 4. At the periphery, the lesion shows some endothelial lined capillaries, large histiocytic cells and giant cells (H&E, x225).
**Immunostaining**

Formalin-fixed, paraffin-embedded sections were deparaffinized in xylol and rehydrated with graded concentrations of ethyl alcohol. The sections were then incubated for 30 min at 37°C in 0.1% Trypsin and 0.1% calcium chloride in distilled water at pH 7.8 in an attempt to expose antigenic sites (1). Following Trypsin digestion, the sections were washed in phosphate-buffered saline (PBS) 0.05 M, pH 7.4 for 15 min. The sections were then layered with appropriate dilutions of 4 specific primary antisera (Table 1). Incubation with the primary antisera was carried out in an humidity chamber overnight at 4°C. The sections were washed with 2 changes of PBS for 30 min at 4°C. Secondary antibodies were applied in appropriate dilutions and incubated for 3 h at room temperature. The sections were again washed with PBS for 30 min. The sites of localization of the peroxidase label were then identified by formation of red color utilizing 4-amino-3-ethyl-carbazole (AEC) which gives a distinct and stable red color. Appropriate

**Table 1. Aneurysmal angiomatoid fibrous histiocytoma.**

<table>
<thead>
<tr>
<th>Monoclonal antibody</th>
<th>Tissue specificity</th>
<th>Source</th>
<th>Result</th>
</tr>
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<tbody>
<tr>
<td>Co-Mo-2</td>
<td>Monocytes</td>
<td>Hybritech-Calif.</td>
<td>+</td>
</tr>
<tr>
<td>Factor VIII</td>
<td>Endothelial Cells</td>
<td>Dako-Calif.</td>
<td>-</td>
</tr>
<tr>
<td>Vimentin</td>
<td>Intermediate Filaments</td>
<td>Lab System-Illinois</td>
<td>-</td>
</tr>
<tr>
<td>Lysozyme</td>
<td>Histiocytic Cells</td>
<td>Miles-Indiana</td>
<td>-</td>
</tr>
</tbody>
</table>
positive controls were stained concurrently. Negative controls consisted of a substitution of non-immune normal serum for the primary antibody. For Factor VIII-Related Antigen Peroxidase-antiperoxidase with the Dako kit was used.

Co-Mo-2, a monoclonal antimonocyte antibody (2) stained many of the tumor cells within the solid portion positively (Table 1). The staining pattern was predominantly cytoplasmic and, in part, finely granular. Factor VIII-related antigen (3) for endothelial cells showed no positive staining cells within the solid portion of the growth. Positive endothelial cell staining occurred in some vascular channels present at the periphery of the lesion. Vimentin for intermediate filaments of fibroblasts (4) showed no positive staining. Lack of staining of vascular endothelium and smooth muscles in this lesion suggest that this antibody did not work in the available material, perhaps because of prior formalin fixation. Lysozyme for histiocytic cells (5) showed no positive cells.

Comments

The term "aneurysmal sclerosing hemangioma" of the skin was first applied by Hairston and Reed (6) when they described 3 patients with large tumors near the knee. Histologically, the lesions showed solid cellular areas and areas rich in vascular elements. The solid portion consisted in part of massive proliferation of foamy histiocytes and occasional multinucleate giant cells. Other areas were populated with spindle and stellate cells associated with collagen bundles. The vascular component showed blood-filled spaces which lacked an endothelial lining and were considered to be the result of hemorrhagic necrosis. Telangiectatic endothelial lined blood vessels were present in the adjacent areas. In 1981, under the title "Aneurysmal (angiomatoid) Fibrous Histiocytoma of the Skin", Santa Cruz and Kyriakos (7) reported a series of 17 cases with pigmented cutaneous nodular lesions located over the extremities and trunk measuring 0.6 to 4.0 cm in diameter. Histologically, the lesions were characterized by the presence of large blood-filled spaces lacking endothelial lining and surrounded by a proliferation of histiocytes, some with foamy cytoplasm or containing hemosiderin pigment granules. Other areas showed proliferation of spindle-shaped fibroblasts and areas of extravasation of erythrocytes resembling Kaposi's sarcoma. A malignant form of angiomatous fibrous histiocytoma was reported by Enzinger (8) in a series of 41 patients, the growths occurring primarily over the extremities of children and young adults. Histologically, the tumors showed solid areas of proliferation of fibroblasts and histiocytic cells, some containing hemosiderin or lipid, areas of hemorrhage, and hemorrhagic cystic formation. Of 24 patients followed, 11 showed recurrence and 5 had metastasis.

The lesion in our case was subcutaneous and appeared to develop, in part, within an aneurysmal blood vessel. The growth showed solid cellular areas and areas with a vascular component. The solid areas consisted of proliferating histiocytes, some with foamy cytoplasm or containing hemosiderin pigment granules. There were also areas showing proliferation of spindle-shaped cells and extravasation of erythrocytes, giving the initial impression of Kaposi's sarcoma. The immunostaining revealed negative Factor VIII for endothelial cells in the solid portion of the lesion eliminating the possibility of Kaposi's sarcoma. The majority of the tumor cells gave a positive reaction with Co-Mo-2, suggesting proliferation of cells of the monocytic series. The lesion was surgically excised; there has been no evidence of local recurrence in the past 18 months.

Aneurysmal (angiomatoid) fibrous histiocyt-
toma may occur in young adults and should be considered in the differential diagnosis of nodular vascular lesions and Kaposi's sarcoma.

References


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