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Orbital Hemangiopericytoma/Solitary Fibrous Tumor in Childhood

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**Abstract:**

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*Sara F. Ribeiro, M.D.*, *Fernando Chahud, M.D.*, and *Antonio Augusto V. Cruz, M.D.*

**Abstract:** A 12-year-old girl had a 6-year history of a large soft-tissue mass in her left orbit. The tumor biopsy was previously performed elsewhere when she was 7 years old, but no treatment was offered at that time. Later, the tumor was completely excised, and histologic examination revealed a mesenchymal neoplasia with typical hemangiopericytoma features. At 9 months of follow up, no evidence of local recurrence or metastasis was seen.

In 1942, Stout and Murray described a mesenchymal neoplasm that they named hemangiopericytoma (HPC) because they considered that the tumor cells derived from pericytes. According to the authors, HPC is a tumor composed of groups of round to spindle cells surrounding endothelium-lined branching vessels that show a staghorn appearance.

Orbital tumors with the histologic appearance described by Stout and Murray are extremely rare in children. In the present report, we describe the case of a girl with a 6-year history of a large orbital lesion, which had all the histologic features considered to be typical of HPC, and we review the literature on this type of orbital neoplasm in children.

**CASE REPORT**

A 12-year-old girl was seen with a painless, large mass in the inferior aspect of the left orbit. The left eye was clearly displaced upward, but eye motility was normal (Fig. 1, top). MRI of the orbits showed a well-defined, large, orbital mass, in the left inferior orbital quadrant, without bone erosion (Fig. 2).

The tumor was completely excised through an inferior eyelid crease approach to the orbit. On gross examination, the tissue fragment measured 3.0 × 2.5 × 1.0 cm and was partially covered by adipose tissue of normal appearance. The cut surfaces showed a grossly round, well-delimited, firm, yellowish brown lesion measuring 2.0 cm in diameter. Microscopic examination revealed a multilobulated tumor with distinct small perivascular nodules outside the main tumor mass (Fig. 3, middle). A ramifying vasculature in a staghorn vascular pattern was seen, with partially hyalinized vessels (Fig. 3, top). The cells around the vessels were mostly round and showed fine chromatin without atypia. Collagen deposition, significant mitotic activity, or necrosis was not found. There was one mitotic figure per 10 high-power fields. Focally, there was an area of myxoid appearance. The reticulin stain revealed a meshwork of reticulin surrounding the neoplastic cells. The immunohistochemical profile of the lesion showed positivity of the neoplastic cells for vimentin, CD34 (Fig. 3, bottom), smooth muscle actin (1A4), HHF-35, and calponin. The surgical margins were free of tumor. No complications occurred after the surgical procedure. Nine months after excision, no evidence of local recurrence or metastases was found.

**DISCUSSION**

Pericytes are elongated periendothelial cells found in almost all capillaries and in small venules and arterioles. They have myoid features reflected in numerous cytoplasmic microfilaments resembling actin- and myosin-containing muscle fibers and immunopositivity for α-smooth muscle actin.

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this reason, pathologists who evaluate CD34+ tumors consider the presence of a meshwork of pericellular reticulin and poorly differentiated myofibrils within the neoplastic stromal cells to be important histologic features for the diagnosis of HPC. However, over the years, it became clear that a variety of different tumors might display the histologic characteristics that once were considered to be typical of HPC. Solitary fibrous tumor (STF) is the most cited example and, although some morphologic criteria are used to differentiate HPC from STF (Table), a general trend is now to consider both tumors as just one entity. In some cases, it is almost impossible to distinguish between STF and HPC morphologically and/or immunohistochemically.

Because the precise line of differentiation shown by HPC/SFT is still unclear, our case could be classified as HPC/SFT (cellular form), even though it presented markers that indicate myoid differentiation. These mesenchymal neoplasms are extremely rare in childhood. We found only 9 tumors classified as HPC and 2, as STF in patients younger than 10 years. They usually have a benign course, even though those that have cytologic atypia, necrosis, and 4 or more mitotic figures per 10 high-power fields can be locally aggressive or metastasize. In addition, some tumors can cause life-threatening hemorrhages.

FIG. 1. Clinical aspect of the patient. Top, Preoperative presentation. Large mass protruding through the inner aspect of the lower eyelid. Notice the scar of a previous biopsy and the vertical eye dystopia. Bottom, Good result after an infraciliary incision, with a slight residual vertical eye dystopia.

FIG. 2. Magnetic resonance imaging. Left, Axial T2-weighted slice showing a multilobulated large mass on the orbital floor. Right, The lesion is intensely enhanced on T1-weighted coronal slice after contrast administration.

FIG. 3. Top, Dense cellularity and staghorn vascular pattern in hemangiopericytoma (hematoxylin-eosin, ×100); middle, multilobulated tumor with distinct perivascular small nodules outside the main tumor mass (hematoxylin-eosin, ×25); bottom, diffuse positivity for CD34 in endothelial cells and cells surrounding blood vessels (immunohistochemistry, ×200).
Criteria used for the histopathologic differentiation between solitary fibrous tumors and hemangiopericytomas

<table>
<thead>
<tr>
<th>Features</th>
<th>Solitary fibrous tumors</th>
<th>Hemangiopericytomas</th>
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<tr>
<td>Stroma</td>
<td>Collagenous +++</td>
<td>Collagenous +++</td>
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<td></td>
<td>Cellularity +/-</td>
<td>Cellularity +/-</td>
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<tr>
<td>Spindle cells</td>
<td>++++</td>
<td>Spindle cells ++++</td>
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<td>Round cells</td>
<td>++++</td>
<td>Round cells +++</td>
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<tr>
<td>Vasculature</td>
<td>Staghorn pattern</td>
<td>Staghorn pattern</td>
</tr>
<tr>
<td>CD34 +</td>
<td>Virtually all cases (vessels and stromal cells)</td>
<td>Most cases (vessels and stromal cells)</td>
</tr>
<tr>
<td>Vimentin</td>
<td>++++</td>
<td>++++</td>
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<tr>
<td>Actin</td>
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<td>Smooth muscle actin</td>
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<td>CD99</td>
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REFERENCES


Plasmacytoma Associated With Canaliculitis

Jae Young You, M.D.†‡, Michael L. Glassman, M.D., F.A.C.S.†, Steven A. McCormick, M.D.*‡, and Tatyana Milman, M.D.*‡‡

Abstract: Plasmacytomas are plasma cell neoplasms that rarely involve ocular adnexal tissues as a primary lesion or secondary manifestation of plasma cell myeloma. Only one case of plasmacytoma involving the lacrimal drainage system, to our knowledge, is described in the literature. The clinical presentation of ocular adnexal primary plasmacytoma typically relates to its mass effect. In this clinicopathologic report, we describe an unusual presentation of primary plasmacytoma of the lacrimal canaliculus as infectious canaliculitis.

Extradural plasmacytoma (EMP) is malignancy of plasma cell origin in soft tissue. EMP can occur as a solitary mass (primary EMP) or as a secondary manifestation of systemic plasma cell neoplasia such as multiple myeloma.1 Ophthalmic involvement by EMP is rare, constituting only up to 6% of ocular lymphoproliferative lesions.2 Data on the ophthalmologic manifestations of primary EMP are limited to case reports, in which EMP is typically described as an indolent mass, localized to the orbit or conjunctiva.1 In this case report, we describe an unusual presentation of primary EMP associated with infectious canaliculitis.

CASE REPORT

A 78-year-old man presented with a 3-year history of tearing, redness, and purulent discharge emanating from the left lower punctum. Examination of the affected region failed to reveal palpable mass in the lacrimal sac area or obstruction on irrigation of the left upper punctum. Thus, clinical findings were interpreted as consistent with infectious canaliculitis (Fig. 1). Incision and drainage of the left lower canaliculus were performed. Upon opening the canaliculus, examination revealed an intracanalicular tumor, which was red and polypoid in nature, and numerous luminal white-yellow, large concretions. The concretions were removed, and the mass was debulked, with preservation of the canaliculus, which was patent on saline irrigation. Following pathologic diagnosis of plasmacytoma, the patient underwent oncologic work-up for plasma cell proliferative and lymphoproliferative disease, which was negative. The patient underwent a total dose of 41 Gy of external beam radiotherapy to the affected area while being maintained on topical lubricating and corticosteroid drops. Six months later, the patient has no clinical evidence of recurrent disease, as evidenced by lack of epiphora, lacrimal system obstruction, palpable mass, or recurrence of canaliculitis. Histopathologic evaluation of the biopsied tissue revealed a collection of mildly atypical plasma cells in the canalicular stroma (Fig. 2). Immunohistochemical evaluation showed that the atypical cells expressed plasma cell marker CD138 and weakly

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