Intracranial, Extradural, Hemangiopericytoma in a Neonate

Abstract

Intracranial infantile hemangiopericytoma (HPC) is a rare, sparsely documented neoplasm with a relatively favorable prognosis than its adult counterpart. We describe a neonatal extradural, intracranial, infantile HPC managed with near-total excision.

Keywords: Extradural, hemangiopericytoma, intracranial, neonate, tumor, vascular

Introduction

Hemangiopericytoma (HPC) is an uncommon, highly vascular soft-tissue tumor[1] earlier believed to arise from the pericytes of Zimmerman. Currently, a fibroblastic origin is accepted, and it is classified under fibroblastic/myofibroblast tumors.[2] HPC may be intracranial or peripheral and occurs as infantile and adult forms.

Intracranial HPC is a distinct entity; only 12 cases have been reported in children. The rarity and heterogeneity of this tumor makes management difficult. We report a neonate with intracranial HPC and review the sparse literature.

Case Report

A term, male, 2.4 kg neonate presented with a gradually progressive swelling on the left side of the face since birth. He was born by cesarean section to a 26-year-old primigravida mother with preeclampsia. The 10 cm × 12 cm mass [Figure 1a] was spread over the left upper face and temporal scalp deforming the left palpebral fissure and caused a left eye watery discharge. The overlying skin was stretched and shiny with engorged veins. It had well-defined margins, bosselated surface, and variegated consistency. The swelling was nonpulsatile, carotid pulsations were unremarkable, and the anterior fontanelle was soft. Although the globe of the left eye was distorted, both fundi were normal. A provisional diagnosis of a vascular lesion/malformation or neuroblastoma was made.

Initial laboratory investigations (complete blood counts, urinary catecholamines, and serum alpha fetoprotein) were normal. Computed tomography [Figure 1b and c] showed a large calvarial soft-tissue lesion in the left temporal and adjacent frontoparietal regions with extracranial and intracranial components. There was heterogeneous enhancement and central necrosis, but no calcification. The lesion was extradural with no obvious brain parenchymal invasion; it had minimal extensions into the ipsilateral orbit (through the lateral wall), masticator space, buccal space, parotid space, and upper neck with erosion of the adjacent mandible. The arterial phase showed few twigs from the left external carotid artery (ECA) supplying the mass.

A preoperative diagnosis of a moderately vascular, predominantly extradural neoplasm was made.

At exploration, a well-defined, 10 cm × 12 cm vascular, extradural, variegated mass was excised from the left temporoparietal region. There was a corresponding bony defect with attenuation of the marginal bone. The overlying skin was stretched and shiny with engorged veins. It had well-defined margins, bosselated surface, and variegated consistency. The swelling was nonpulsatile, carotid pulsations were unremarkable, and the anterior fontanelle was soft. Although the globe of the left eye was distorted, both fundi were normal. A provisional diagnosis of a vascular lesion/malformation or neuroblastoma was made.

Grossly, the mass was well circumscribed; it had a homogeneous, grayish-white cut surface with areas of hemorrhage.


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and cystic degeneration. Microscopy [Figure 2] revealed a well-circumscribed, highly cellular mass. Cells were arranged in diffuse sheets with numerous interspersed staghorn-shaped blood vessels [Figure 2a]. Individual tumor cells were monomorphic with oval-to-spindle-shaped nuclei, nuclear grooving, bland nuclear chromatin, and scant-to-moderate amount of cytoplasm [Figure 2b]. The mitoses were largely few, occasional patches showed brisk mitotic activity (1–3/high-power field). Erythroid colonies, myeloid precursors, and occasional megakaryocytes were scattered within the tumor cells at multiple foci suggesting extramedullary hematopoiesis [Figure 2c]. In addition, there were areas of hemorrhage, collections of siderophages, and cystic degeneration. Immunohistochemistry for vimentin (cytoplasmic) and CD99 (membranocyttoplasmic) [Figure 2d] showed diffuse strong positivity. CD34 immunostain was positive (membranous) in a subset of tumor cells [Figure 2e]. Smooth muscle actin positivity was patchy cytoplasmic, and there was diffuse nuclear STAT6 positivity [Figure 2f]. Leukocyte common antigen (LCA), glial fibrillary acidic protein, CD31, pan-cytokeratin, CD1a, Bcl2, desmin, S100, and Myeloperoxidase (MPO) were negative in the tumor cells. LCA and MPO highlighted the interspersed hematopoietic cells in the background and CD31 highlighted the interspersed vessels. A diagnosis of infantile intracranial HPC was rendered combining the histomorphology and immunohistochemistry.

Figure 1: Clinical photograph (a) of the protuberant temporal mass distorting the left palpebral fissure. Contrast-enhanced computed tomography axial view (b) and coronal reformatted image (c) showing the large, heterogeneously enhancing calvarial mass. Postoperative appearance (d) at 6-month follow-up. Contrast-enhanced computed tomography axial view (e) and coronal reformatted image (f) showing enhancing residual extracranial component (black arrow) and postoperative cystic cavity (star) in the left temporal region and infratemporal fossa.

Figure 2: (a) Microscopy showed a cellular tumor in diffuse sheet with interspersed staghorn-shaped blood vessel (H and E, ×100) (b) The individual cells showed spindle-shaped nuclei, nuclear grooving, and bland chromatin (H and E, ×400) (c) Interspersed were erythroid colonies (black arrow) and megakaryocytes (blue arrow) (H and E, ×100) (d-f) Positive immunohistochemistry for CD99 (membranocyttoplasmic, ×200, d), CD34 (membranous, ×200, e), and STAT6 (nuclear, ×400, f)
Intracranial hemangiopericytoma in a neonate

Pediatric HPC commonly occurs in the soft tissues of the lower extremities, an intracranial location is more likely in adults. Intracranial HPC is commonly seen in the fifth to sixth decades of life; of the 5%–10% cases occurring in childhood, 40% occur in the 1st year of life. Two distinct clinical entities exist in pediatric HPC. Pediatric HPC, a sarcomatous soft-tissue tumor of vascular origin, constitutes only 1% of all vascular tumors. HPC is commonly seen in the fifth to sixth decades of life; of the 5%–10% cases occurring in childhood, 40% occur in the 1st year of life. Two distinct clinical entities exist in pediatric HPC. Pediatric HPC, a sarcomatous soft-tissue tumor of vascular origin, constitutes only 1% of all vascular tumors. HPC is commonly seen in the fifth to sixth decades of life; of the 5%–10% cases occurring in childhood, 40% occur in the 1st year of life. Two distinct clinical entities exist in pediatric HPC. Pediatric HPC, a sarcomatous soft-tissue tumor of vascular origin, constitutes only 1% of all vascular tumors.

Neonatal soft-tissue tumors display large phenotypic variations due to the intrinsic multipotential nature of mesenchymal tissues. HPC, a sarcomatous soft-tissue tumor of vascular origin, constitutes only 1% of all vascular tumors. HPC is commonly seen in the fifth to sixth decades of life; of the 5%–10% cases occurring in childhood, 40% occur in the 1st year of life. Two distinct clinical entities exist in pediatric HPC. Pediatric HPC, a sarcomatous soft-tissue tumor of vascular origin, constitutes only 1% of all vascular tumors.

The neonate made an uneventful recovery. At 9 months postoperative follow-up, he is thriving well.

Table 1: Comparative summary of reported cases of neonatal intracranial hemangiopericytoma

<table>
<thead>
<tr>
<th>Author</th>
<th>Sex</th>
<th>Antenatal diagnosis</th>
<th>Clinical features and location of mass</th>
<th>Management</th>
<th>Histopathology</th>
<th>Reti culin</th>
<th>Vimentin</th>
<th>Immunohistochemistry</th>
<th>F/U and outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peace (1954)</td>
<td>Male</td>
<td>No</td>
<td>Flaccid seizures, bulging fontanelle</td>
<td>Nil</td>
<td>Proliferation of ovoid cells, hemorrhage, dense capillary</td>
<td>Rich</td>
<td>NM</td>
<td>CD34</td>
<td>Death</td>
</tr>
<tr>
<td>Solitaire and Krigman (1964)</td>
<td>Female</td>
<td>No</td>
<td>Stillborn</td>
<td>Nil</td>
<td>Fusiform cells, “mixed hemangiopericytoma and meningeal fibroma”</td>
<td>Rich</td>
<td>NM</td>
<td>NM</td>
<td>Death</td>
</tr>
<tr>
<td>Aouad et al. (1991)</td>
<td>Male</td>
<td>No</td>
<td>Lethargy, tense fontanelle, papilledema</td>
<td>Complete gross resection</td>
<td>Highly proliferative ovoid cells surrounding thin-walled capillaries, extensive necrosis, numerous mitotic figures</td>
<td>Rich</td>
<td>Positive</td>
<td>NM</td>
<td>Negative</td>
</tr>
<tr>
<td>Herzog et al. (1995)</td>
<td>Male</td>
<td>No</td>
<td>Left ptosis</td>
<td>Incomplete resection</td>
<td>Highly cellular lesion, extensive vascular network</td>
<td>Rich</td>
<td>NM</td>
<td>NM</td>
<td>Spontaneous regression by 18 months, well at 27 months</td>
</tr>
<tr>
<td>Cavalheiro et al. (2002)</td>
<td>Male</td>
<td>Yes</td>
<td>Left frontoparietal mass</td>
<td>Complete gross resection</td>
<td>Hypercellular, ovoid cells, hypervascular, necrosis</td>
<td>Rich</td>
<td>Positive</td>
<td>NM</td>
<td>Death</td>
</tr>
<tr>
<td>Sobel et al. (2006)</td>
<td>Male</td>
<td>Yes</td>
<td>Posterior cranial fossa mass</td>
<td>Nil</td>
<td>Highly cellular, extensive vascular network</td>
<td>NM</td>
<td>Positive</td>
<td>Negative</td>
<td></td>
</tr>
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</table>
In the 2016 WHO classification of central nervous system tumors, solitary fibrous tumor (SFT) and HPC are deemed as one entity in the group of mesenchymal, nonmeningothelial tumors. Currently, HPC is considered to be of fibroblastic origin. The WHO classification of tumors of the central nervous system (2016) has taken a unified approach and considered HPC and SFT to be part of the same spectrum with identical molecular features but different phenotypes. SFT shows a “patternless” pattern and is less cellular due to abundant deposition of collagen, whereas HPC shows high cellularity. Both phenotypes show NAB2-STAT6 fusion on molecular testing. This gives rise to an upregulation of STAT6 protein detected by immunohistochemistry, as in the index case. On these lines, we have combined the histomorphology and immunohistochemical features in the index case to render a diagnosis of intracranial HPC.

In conclusion, intracranial, infantile HPC is rare and has a favorable prognosis compared to its adult counterpart. In an extradural form, complete surgical resection is feasible with surveillance of minor residua.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**References**


