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Case Report

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*

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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 spreads 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 small, diffuse, firm noncontiguous mass in the infratemporal fossa with a separate investment was left undisturbed for a staged management after review of the histopathology. There was no dural tear, and the skin flaps were closed over the defect.

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 (membranocytoplasmic) [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.

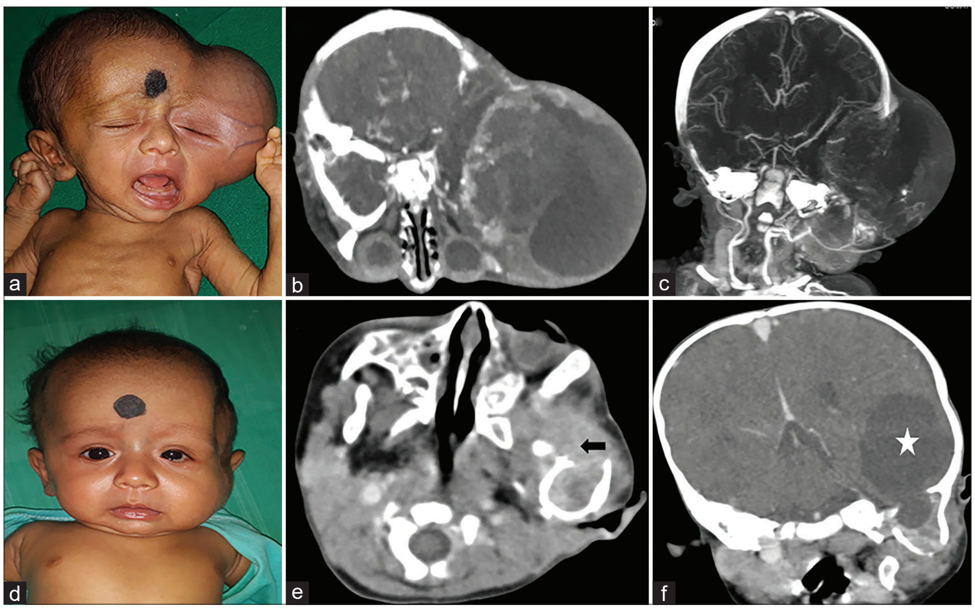

Figure 1: Clinical photograph (a) of the protuberant temporal mass distorting the left palpebral fissure. Contrast-enhanced computed tomography axial view (b) and coronal reformatted image (c) showing the large, heterogeneously enhancing calvarial mass. Postoperative appearance (d) at 6-month follow-up. 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

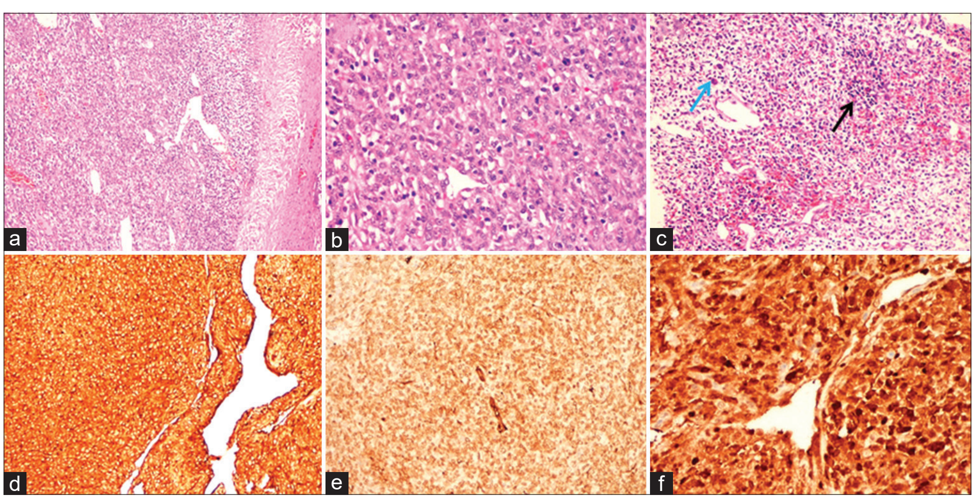

Figure 2: (a) Microscopy showed a cellular tumor in diffuse sheet with interspersed staghorn-shaped blood vessel (H and E, ×100) (b) The individual cells showed spindle-shaped nuclei, nuclear grooving, and bland chromatin (H and E, ×400) (c) Interspersed were erythroid colonies (black arrow) and megakaryocytes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

The neonate made an uneventful recovery. At 9-month postoperative follow-up, he is thriving well [Figure 1d]. Contrast-enhanced computed tomography shows no intracranial component; the residual infratemporal mass [Figure 1e and f] is static and under surveillance. In the absence of a clear guideline for chemotherapy, an informed decision was made after parental counseling to follow-up closely.

Discussion

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.[3] 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 cases beyond infancy behave like adult HPC; they are common in extremities, aggressive, and respond poorly to chemotherapy. In contrast, infantile HPCs are histologically similar tumors that are less aggressive, respond better to chemotherapy, show spontaneous regression, and have an overall favorable prognosis.[1,2,4]

Pediatric infantile HPC commonly occurs in the soft tissues of the lower extremities; an intraoral location is more likely than in adults.[5] Intracranial location has been reported in only 12 cases in the English published literature; seven of these are neonates [Table 1]. Two were diagnosed antenatally, one was stillborn, and three had died without treatment.[1] Herzog *et al*.[6] describe two cases of intracranial HPC, one of whom had a left temporal mass with ophthalmoplegia akin to the case described here.

Surgical excision is the mainstay of treatment in infantile HPC. The timing of surgery has varied from 2 days to 18 months. Although gross total resection is desirable, it may be technically difficult. We have removed the bulky mass and left a smaller residuum in the infratemporal fossa with a separate investment for a subsequent procedure. Hypervascularity is common; despite the surface location of the tumors, its vascular supply is generally from the internal carotid artery or its branches. In the described case, the dural branches of the ECA supplied the mass. Herzog *et al*.[6] describe a neonate who underwent a partial resection of a left infratemporal mass and had a residual left behind in the temporal lobe to avoid further intraoperative blood loss. Despite no postoperative therapy, the mass had regressed at 18-month follow-up. Spontaneous regression of the tumor has also been reported in other sites.[7,8] There is no standard adjuvant chemotherapy, however spontaneous regression has been documented after subtotal resection. Therefore, we have opted to monitor the static residua further and reserve complete surgical resection for persistence or progress.

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

Author (year)	Sex	Antenatal diagnosis	Clinical features and location of mass	Management	Histopathology	Reticulin	Immunohistochemistry					F/U and outcome
							Vimentin	CD34	GFAP	Desmin	S100	
Peace (1954)	Male	No	Flaccid seizures, bulging fontanelle Right cerebral mass	Nil	Proliferation of ovoid cells, hemorrhage, dense capillary network	Rich	NM	NM	NM	NM	NM	Death
Solitare and Krigman (1964)	Female	No	Stillborn Right middle cranial fossa mass	Nil	Fusiform cells, "mixed hemangiopericytoma and meningeal fibroma"	Rich	NM	NM	NM	NM	NM	Death
Aouad *et al.* (1991)	Male	No	Lethargy, tense fontanelle, papilledema Right cerebral	Complete gross resection	Spindle-shaped cells, numerous thin-walled vascular channels, necrosis	Rich	Positive	NM	Negative	NM	Negative	Well at 5 months
Herzog *et al.* (1995)	Male	No	Left ptosis Left anterior temporal fossa	Incomplete resection	Highly proliferative ovoid cells surrounding thin-walled capillaries, extensive necrosis, numerous mitotic figures	Rich	Positive	NM	Negative	NM	Negative	Spontaneous regression by 18 months, well at 27 months
Cavalheiro *et al.* (2002)	Male	Yes	Left frontoparietal mass	Complete gross resection	Highly cellular lesion, extensive vascular network	Rich	NM	Positive	NM	NM	NM	Well at 24 months
Sobel *et al.* (2006)	Male	Yes	Posterior cranial fossa mass	Nil	Hypercellular, ovoid cells, hypervascular, necrosis	NM	Positive	Positive	Negative	Negative	Negative	Death

Contd...

Table 1: Contd...

Author (year)	Sex	Antenatal diagnosis	Clinical features and location of mass	Management	Histopathology	Reticulin	Immunohistochemistry					F/U and outcome
Kerl *et al*. (2011)	Female	No	Dehiscence of sagittal fissure Left occipitomental mass	Eight cycles chemo-VAC, complete resection at 18 months	Highly vascular, cellular, necrosis	NM	NM	Positive	NM	NM	NM	Well, at?
Present case (2020)	Male	No	Left temporofacial disfiguring mass	Incomplete resection	Highly cellular, staghorn-shaped blood vessels	ND	Positive	Positive	Negative	Negative	Negative	Static residual, at 9 months

NM – Not mentioned; ND – Not done; GFAP – Glial fibrillary acidic protein; F/U – Follow-up; VAC – Vincristine, Actinomycin D, Cyclophosphamide

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.[9] 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.[9] 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.

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