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CASE REPORT - PEDIATRICS

Giant intracranial hemangioma in a neonate

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Abstract In this report we detail the case of an infant presenting with a giant intracranial congenital hemangioma and describe the clinical features and surgical management. Congenital hemangiomas are benign vascular tumors that typically present as skin lesions in neonates and infants. On rare occasions they present as intracranial tumors. The possibility that these tumors may undergo spontaneous regression poses a treatment dilemma.

Keywords Hemangioma · Pediatric brain tumor · Vascular tumor · Vascular disorders

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Introduction

Intracranial vascular tumors presenting in the first year of life are a rare entity that pose special diagnostic and treatment challenges. Large intracranial lesions in this age group, albeit uncommon, are more likely to be neoplastic with primitive neuroectodermal tumors, astrocytoma and choroid plexus papilloma being the most frequently encountered histological subtypes [13]. The most common clinical presentation, regardless of the underlying pathology, is with signs of raised intracranial pressure and/or enlarging head circumference [13]. In addition to the non-specific clinical features CT, MRI and angiography may not help in distinguishing these lesions.

Hemangiomas are benign vascular tumors that typically involve the skin of the head, neck and chest. They constitute the most common tumors of childhood but rarely affect the central nervous system (CNS) [8]. They may continue to grow during infancy but can also spontaneously regress. We report the rare occurrence of a very large and entirely intracranial congenital hemangioma in a neonate.

Case report

A full-term infant boy born to a 34-year-old woman presented with a rapidly expanding head circumference, irritability and seizures two weeks following an uneventful birth. Signs of raised intracranial pressure, including a tense anterior fontanelle, distended scalp veins, enlarged head circumference, and reduced eye opening, were noted on examination.

CT and MRI demonstrated well-defined enhancing lobulated tumor that appeared to be located extra-axially in the left middle fossa (Fig. 1). The lesion was associated with a very large cyst that occupied much of the left

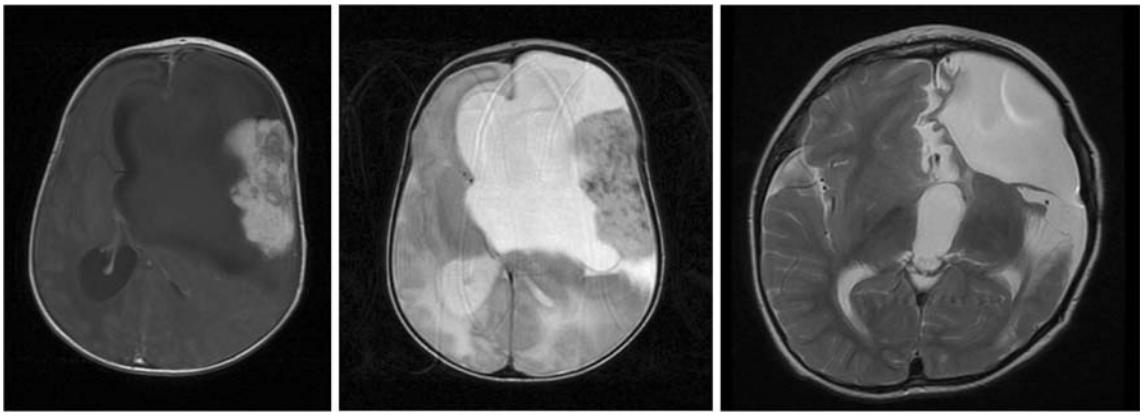

Fig. 1 Preoperative MRI imaging. T1-weighted with gadolinium contrast (*Left*) and T2-weighted (*Middle*) MRI demonstrating an avidly enhancing mass with several flow voids within the solid component associated with a large cystic component occupying much of the left hemisphere. Post-operative T2-weighted MRI (*Right*) demonstrates no recurrence of the lesion but a large cyst occupying the anterior half of the left hemisphere

hemisphere resulting in considerable displacement of midline structures.

An open biopsy of the solid component and decompression of the cyst were performed urgently. As expected the tumor was highly vascular. Histology suggested a mixed capillary and cavernous hemangioma with dilated capillary-sized endothelium-lined vessels on microscopy.

In view of continued expansion of our patient's head circumference due to cyst enlargement a surgical resection was carried out three weeks after the biopsy. At surgery a chocolate-brown tumor covered superiorly by thin brain mantle and with a dissectible plane was encountered. Numerous high-flow vascular pedicles of cortical arteries and veins associated with the lesion were coagulated and divided to allow the complete resection of a 6×5×2.5 cm encapsulated tumor.

Microscopy revealed a thin fibrous limiting pseudocapsule with a marginal large arterio-venous leash. Much of the lesion consisted of dilated capillary-sized vessels with flattened endothelial cells. The dilated vessels were not as large or as extensive as those observed in cavernous malformations. One short stretch of brain-tumor interface showed shallow capillary intrusion into the brain but, for the most part, brain and lesion had a non-infiltrative 'pushing' border-zone of connective tissue (Fig. 2). There were multiple foci of fresh hemorrhage, organizing clot, vascular thrombosis and mineralization. The vascular nature of the tumor was confirmed by CD31 and CD34 staining of the endothelium. Positive GLUT1 staining, which distinguishes infantile hemangiomas from congenital hemangiomas [8], was largely absent.

Were the lesion to have been extra-cranial, it would best fit with either a rapidly-involuting congenital hemangioma (RICH) or non-involuting congenital hemangioma (NICH). We interpreted the areas of thrombosis and focal interstitial fibrosis as involutional change, thus, favoring the former.

Postoperative recovery was uneventful with satisfactory neurological progress but with delayed attainment of developmental milestones. Continued anti-epileptic medication and a cysto-peritoneal shunt were required. Post-operative imaging performed two years after surgery demonstrates a residual cyst but no recurrence (Fig. 1).

Discussion

Large intracranial lesions are rare in the neonate. A primitive neuroectodermal tumor or astrocytoma would be considered the most likely diagnosis [13]. The discovery of a hemangioma is an unexpected but important finding given the potential for curative treatment and also the possibility that some of these lesions may spontaneously regress.

Hemangiomas are common vascular tumors in children. They occur on the skin of the head, neck, and chest, but rarely occur intracranially [8]. When they do occur in the CNS it tends to be in extra-axial locations, such as the dural sinuses or in the orbit, and often in association with skin lesions. [2, 7, 12, 14, 15] A solitary hemangioma is a rare finding with only a few reported in the literature and certainly none of this size.

Viswanathan et al., reported a large series of nine intracranial hemangiomas in neonates and infants [15]. Unlike our case, none of their patients had solitary CNS disease. In the majority, imaging actually revealed a connection between cutaneous disease and the intracranial lesion. None of their patients had hemangiomas that invaded the brain or showed significant mass effect. However, prominent intralesional flow voids on MRI were noted in several of their cases in keeping with the high flow vascular nature of the hemangioma in our case.

Histologically, hemangiomas are benign vascular tumors featuring clusters of thin walled endothelial-lined spaces filled with blood and/or thrombus at varying stages of organization [1, 11]. They share common histological features with intra-axial cavernous malformations that are more commonly

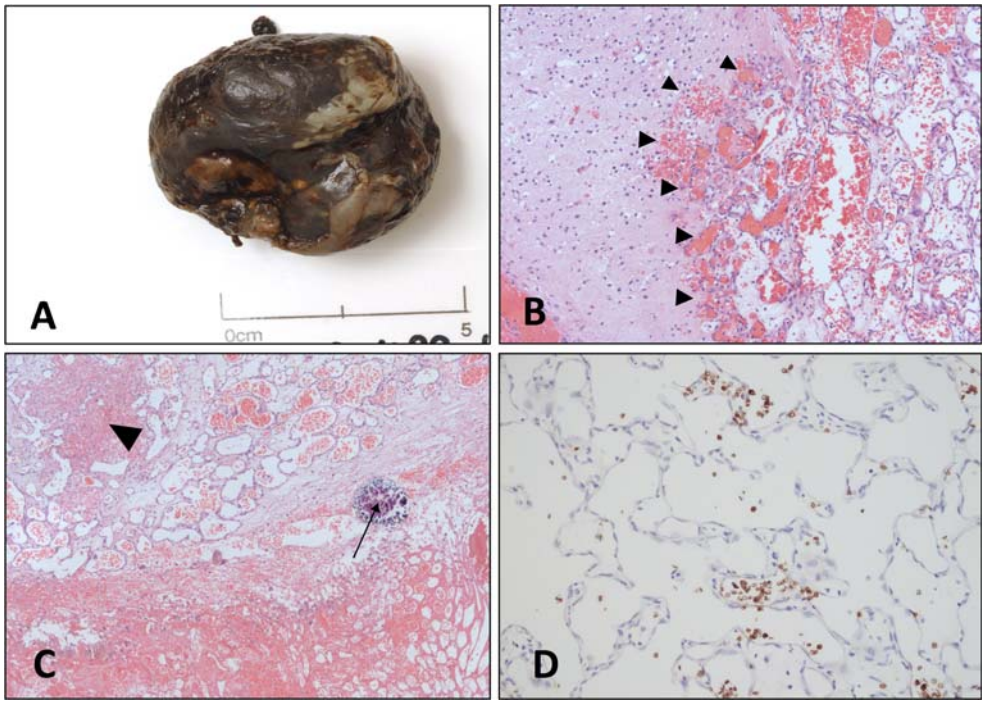

Fig. 2 Macroscopic and microscopic histological specimens. Gross specimen (**a**) and microscopic specimens with hematoxylin and eosin stain (**b** and **c**) and GLUT1 stain (**d**). **a** The macroscopic appearances of the tumor were of a 6×5×2.5 cm chocolate-brown, spongy ovoid. **b** Much of the lesion comprised dilated capillary-sized vessels with flattened endothelial cells. One short stretch of interface showed shallow capillary intrusion into the brain (*arrowheads*) but, for the most part, brain and lesion had a non-infiltrative 'pushing' narrow border-zone of connective tissue. **c** There were multiple foci of fresh hemorrhage and several instances of vascular thrombosis (*arrow head*), interstitial fibrosis and mineralization (*small arrow*). **d** Overall, the endothelium largely stained negative for GLUT1

described in adults [11]. The use of non-standardized nomenclature for intracranial vascular lesions has lead to some confusion regarding their classification. Capillary hemangioma, cavernous hemangioma, cavernous angioma, cavernous malformation, and cavernomas are all terms that have been used to describe histologically similar lesions in the neonate, although there does appear to be at least two distinct entities. Cavernous malformations or cavernomas are considered as malformations or hamartomatous lesions, whereas hemangiomas can be considered neoplastic with the ability to grow and compress adjacent structures [4, 8]. The dilated endothelial-lined spaces of our hemangioma were not as cavernous or as extensive as those typically seen in cavernous malformations. Other features of hemangiomas that can distinguish them from cavernous malformations are their extra-axial location including the dura mater [10], basal cisterns [15], ventricles [6] and cavernous sinus [4], association with cutaneous lesions [15], potential to grow and regress spontaneously over time [8, 14], the presence of a capsule or pseudocapsule [4], and their behavior as high-flow vascular tumors that may hemorrhage profusely during resection [6, 12].

Extra-cranial hemangiomas are classified as 'congenital' or 'infantile' based on growth patterns and immunohistochemal staining for GLUT1. Our lesion most resembled a congenital hemangioma, specifically a rapidly-involuting congenital hemangioma (RICH), based on negative staining for GLUT1 and areas of thrombosis and interstitial fibrosis that we interpreted as involution [8].

Our case typifies the clinical presentation of large tumors in this age group that present with signs of raised intracranial pressure. Seizures, neurological deficits and irritability are also common features [3, 5, 9, 11, 16]. The rather non-specific features of an enhancing lobulated tumor found on CT and MRI were not helpful in determining a diagnosis. Occasionally, CT and MRI may reveal areas of focal calcification and evidence of recent hemorrhage that would be more in keeping with a vascular lesion [3, 10, 11]. Similarly, angiography does not appear to be useful, only occasionally demonstrating a vascular blush [3, 16].

Given the rarity of neonatal intracranial hemangioma, it is not surprising that the natural history of these lesions is unclear. Certainly analogous extra-cranial lesions may actually involute spontaneously after the neonatal period [11]. In patients with cutaneous hemangioma who are found to have an intracranial lesion, it may be appropriate to watch for regression. Tortori-Donati et al., were able to follow small meningeally-based lesions in four infants with cutaneous hemangiomas [14]. In two cases regression of the intracranial disease was observed in parallel to regression of the cutaneous disease. In our case the lack of cutaneous disease, the large size and the increasing mass effect caused by the hemangioma necessitated surgical resection. Previous reports of neonatal

hemangioma, like our case, have also reported satisfactory surgical resection that is curative with low morbidity [7, 11]. Failure to achieve total resection may result in a poor outcome due to the risk of early hemorrhage from residual hemangioma [11].

Conclusion

Neonatal hemangioma is a rare lesion that may be difficult to distinguish from more common tumors. Histologically, they are benign tumors that can grow to large sizes as demonstrated by our case. Complete surgical resection when achievable is safe and curative. There may be the option, however, to watch for spontaneous regression if the lesion is small and associated with cutaneous disease.

Conflicts of interest None.

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