Congenital cystic supratentorial hemangioblastoma

Case report

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SUPRATENTORIAL hemangioblastomas are exceedingly rare tumors. The first case was described by Bielschowsky in 1902; since then approximately 83 cases have appeared in the literature. Of these, only one case has been reported in the first year of life; however, no details of that case were presented, including the exact age of the patient at presentation. The following report involves a case of supratentorial hemangioblastoma occurring in a 3-week-old infant, which we believe is the youngest age at which this tumor has been described.

Case Report

This 3-week-old baby boy was referred for evaluation of a head circumference that had been gradually expanding since birth. The child was the product of an uncomplicated pregnancy. An ultrasound study at 28 weeks of gestation had shown no abnormality.

Examination. Physical examination revealed an alert but irritable child. The fontanels were tense and bulging. The veins of the forehead were prominent and the child was noted to have “sunset” eyes; otherwise, he was neurologically intact. The hematocrit was within normal limits for his age.

Computerized tomography (CT) of the head revealed a large, loculated cystic mass occupying much of the left hemisphere, with a 2.5-cm inhomogeneous hyperdense mural nodule at its medial aspect. Contralateral ventriculomegaly was also present. The child was placed under general anesthesia and magnetic resonance imaging was performed which confirmed the CT findings (Fig. 1).

Operation. The mass was exposed via a left frontoparietal craniotomy. The tumor appeared cherry-red and highly vascular (Fig. 2). The mass was well circumscribed and no dural attachment was demonstrated; it was cleanly separated from the surrounding gliotic tissue and removed intact. Figure 3 shows the cut surface of the surgical specimen.

Pathological Examination. Histological examination of the surgical specimen revealed numerous thin-walled vascular channels lined with plump and swollen endothelial cells. The capillary structures were separated by numerous ovoid, vacuolated stromal cells. Mitoses and cellular atypia were absent (Fig. 4).

Postoperative Course. The patient’s postoperative course was uneventful, and he was discharged home on the 12th postoperative day. In view of the possible association with von Hippel-Lindau disease, further follow-up studies were undertaken. Results of a detailed funduscopic examination and a bilateral renal ultrasound were both found to be normal. In addition, funduscopic and renal ultrasound examinations were performed on both parents and these results were also without abnormality.
Discussion

Hemangioblastomas are benign tumors occurring exclusively within the neuraxis. They account for 1.5% to 2.5% of all intracranial tumors. These lesions are most commonly found in the cerebellum, and comprise 7% to 12% of all posterior fossa neoplasms. Supratentorial hemangioblastomas, however, are exceedingly rare, with approximately 83 cases reported in the literature to date.

In the pediatric population, supratentorial hemangioblastoma is even rarer. We have found only four previous cases of supratentorial hemangioblastoma occurring in children under 10 years of age, and at least one of these was thought to be an angiolastic meningioma. This distinction is relevant because, in contrast to hemangioblastoma, angiolastic meningioma occurs most frequently in the supratentorial compartment. Congenital tumors are defined as those presenting in the first 60 days of life. Of the four reported cases of supratentorial hemangioblastomas, only one was found in a child under 1 year of age, but the details of that case, including the exact age at presentation, were not presented. In 1936, Bergstrand, et al., described a case of congenital hemangioblastoma, which we believe to be the only other reported case of a hemangioblastoma that was definitely congenital. However, the location of the tumor within the brain (supratentorial vs. infratentorial) was not specified.

Controversy exists in regard to the distinction between a true hemangioblastoma and an angiolastic meningioma. Frequently, the histological picture of both tumors is indistinguishable, with the difference being found mainly in the gross appearance and topographical location. The presence or absence of a dural attachment was suggested by Cushing and Eisenhardt to be the most important factor to consider in distinguishing the two tumor types; however, Russell and Rubinstein stated that a reliable diagnosis could not be based on this feature alone. They noted that angiolastic meningioma is usually a solid tumor attached to the dura, whereas hemangioblastomas are often cystic, with a mural nodule within the wall of the cyst, and lack a dural attachment. These findings, combined with the characteristic gross and histological appearance, provide strong evidence for the separation of hemangioblastoma from angiolastic meningioma.
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tological picture, confirmed the diagnosis of hemangioblastoma in our patient.

Supratentorial hemangioblastomas have been reported to occur as a manifestation of the von Hippel-Landau complex. This disease, however, has not been present in the majority of cases$^7,15$ and was not found in our patient, nor was there a positive family history.

In summary, we believe this to be the first reported case of a congenital supratentorial hemangioblastoma. In addition, we believe this to be only the second reported case of congenital hemangioblastoma occurring at any location within the brain. We stress the importance of deferring frozen sections in cases of suspected hemangioblastoma, as reported by Ho, et al.$^8$

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References


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