NGIOSARCOMA is a rare malignant neoplasm of the vascular endothelium that occurs mainly in the cutaneous and soft tissues. Angiosarcoma arising in the CNS has been reported only rarely. Within the CNS, it may present as a pure neoplasm with no other component, as mixed with glioblastoma multiforme, or as a metastatic neoplasm. Clinically, the congenital type of CNS angiosarcoma is particularly rare. To our knowledge, this is only the third report of congenital primary cerebral angiosarcoma.

Case Report

History and Examination. This 30-day-old boy was referred to our department for further treatment for an enlarged head and a 1-week history of projectile vomiting. He had been born after a full-term pregnancy and normal delivery; results of an ultrasonography study obtained at 39 weeks of gestation had been normal. Following admission, results of an abdominal ultrasonography study and physical examination were normal, except for a head circumference of 41 cm (> 2 standard deviations above the mean for his age) and a tense anterior fontanelle. A 2.5-cm heterogeneous hyperdense mass was revealed in the left frontotemporal area near the left insula on CT scanning.

Operation and Postoperative Course. Left frontotemporal craniotomy was performed 4 days after admission. On dissection of a cystic lesion within the left sylvian fissure, the tumor appeared berry-red, highly vascular, and well circumscribed. Total excision of the mass was accomplished without incident. On histopathological examination it was revealed that the neoplasm was composed of ramifying vascular channels of different shapes and calibers, with intraluminal papillary structures lined by plump neoplastic endothelial cells containing hyperchromatic nuclei. The number of mitotic figures was 1.2 high-power fields. Immunohistochemical reaction of the neoplastic cells for endothelium-specific antigens including factor VIII–related antigen, CD31, and CD34 was diffusely positive. With these findings we confirmed the final diagnosis of congenital primary cerebral angiosarcoma. The patient’s postoperative course was uneventful, and he was discharged 2 weeks after the operation. He was in good condition with no sign of recurrence after 11 months; follow-up computerized tomography, magnetic resonance (MR) imaging, and abdominal ultrasonography studies demonstrated no tumor regrowth. The characteristic findings for this tumor on MR imaging, the immunohistochemical findings, and surgical outcome are discussed.

Key Words • cerebral angiosarcoma • congenital tumor

Congenital primary cerebral angiosarcoma

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Abbreviations used in this paper: CNS = central nervous system; CT = computerized tomography; MR = magnetic resonance.
Discussion

Primary intracranial sarcomas are uncommon and are reported to comprise 1 to 2% of primary intracranial neoplasms. Of these tumors, angiosarcoma is particularly rare, accounting for less than 1% of all sarcomas. To our knowledge, only two cases of congenital primary cerebral angiosarcoma have been previously described.

The clinical course of cerebral angiosarcoma is characterized by the rapid onset of symptoms, usually appearing within several weeks before surgery. All reported patients underwent surgical resection of the tumor, with only one receiving postoperative radiation therapy. A primary cerebral angiosarcoma has been described, with its appearance on CT and MR studies reported as a well-demarcated lesion with hemorrhagic characteristics. In our case, despite apparent good demarcation on both radiological studies and visual inspection at the time of surgery, radiological findings in the tumor included severe perifocal edema with cystic formation and marked contrast medium enhancement. The outcome of primary cerebral angiosarcoma is generally poor. A median survival rate of only 8 months has been reported, and investigators were unable to demonstrate any correlation between histological features and biological behavior. However, long-term survivors have occasionally been reported.

Although radiation therapy is reported to be effective for angiosarcoma of the bone or metastatic cerebral angiosarcoma, findings on studies of the effectiveness of radiotherapy for primary cerebral angiosarcomas are still controversial. On the other hand, adjuvant chemotherapy for the treatment of soft-tissue sarcomas is still experimental. In most prospective randomized studies completed so far the researchers have failed to demonstrate clear effectiveness in the treatment of angiosarcomas because of small sample size, poor study design, and other methodological problems. Recently, Antoniadis, et al. presented the case of a 41-year-old patient with primary cerebral angiosarcoma who received adjuvant chemotherapy and radiation after surgical resection of the tumor. The patient was in excellent clinical and neurological condition with no sign of recurrence for 41 months. Our patient underwent surgical resection of his tumor without receiving postoperative radiation therapy or adjuvant chemotherapy, but he was still in good health with no sign of recurrence after 11 months, and follow-up CT, MR, and abdominal ultrasonography studies demonstrated no regrowth.

Microscopically, angiosarcoma is a vascular neoplasm with marked atypia and many mitoses. Immunohistochemical staining with factor VIII, CD31, CD34, and lectin ULEX are essential and useful for differentiating angiosarcoma from meningeal hemangiopericytoma, anaplastic gliomas, metastatic carcinoma, amelanotic melanoma, hemangioblastoma, and various richly vascular sarcomas. In our case, immunohistochemical reaction of the neoplastic cells to endothelium-specific antigens including factor VIII–related antigen, CD31, and CD34 was positive.
diffusely positive. We thus confirmed the final diagnosis of primary cerebral angiosarcoma.

Congenital CNS tumors have been variously considered to be definitely congenital (present at birth), probably congenital (arising within the 1st week of life), and possibly congenital (arising within the 1st month of life). Although our patient was admitted 30 days postnatally, the clinical symptom of projectile vomiting for 1 week began within the defined congenital period. We therefore believe that this patient’s primary cerebral angiosarcoma was congenital in origin. Kirk, et al., presented a 12-day-old girl and Mena, et al., reported on another 2-week-old patient in whom a congenital primary cerebral angiosarcoma was diagnosed; we believe our case is the third report of this disease.

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References

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