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Case reports

Giant cerebellar hemangioma in an infant

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Abstract. A case is reported of an infant with a giant cerebellar vermis hemangioma which was totally removed. The excised specimen contained both cavernous angioma-like and telangiectasia-like portions with intervening cerebellar tissue. The tumor was considered to be a mixture of cavernous hemangioma and telangiectasia.

Key words: Brain tumor – Cavernous hemangioma – Obstructive hydrocephalus – Cerebellar vermis.

According to Russell and Rubinstein [13], intracranial angioma is a vascular hamartoma, a type of congenital anomaly. It is therefore thought to be present prior to birth, but is nevertheless rarely reported in the newborn or infant [4, 11]. In the review by Voigt and Yasargil [16], it is said to have a peak incidence in the 30s, between 20 and 50 years of age. It occurs most frequently in the supratentorial compartment and is rare in the posterior fossa. The incidence of angioma in the cerebellum, including arteriovenous malformation, is 5% to 17% [1, 6, 16]. As far as we know, there is no report of a case with a giant angioma, other than arteriovenous malformation, extensively occupying the cerebellum.

We report the case of a giant vascular anomaly of the cerebellum in an infant; some characteristic points on the histopathology, radiological diagnosis, and treatment are discussed.

Case report

The patient was a 6-month-old female. She was examined because she had had a mild head injury and was admitted with the diagnosis of cerebellar tumor because of a bulging and tense anterior fontanelle, instability of the head for her age, and enlargement of the head. Neurologically, disturbance of the upward gaze, bilateral abducens nerve palsies, hypotonia, and bilateral terminal tremor (probably intension tremor) were observed. The fundi oculi were normal, as were the biochemical and hematologic studies. The family and past histories were noncontributory.

Plain radiographs of the skull showed a linear fracture of the right parietal bone and calcified spots in the median part of the posterior fossa. In the non-enhanced CT scan, aggregations of granules of high density were noted in the median part of the posterior fossa. The third and lateral ventricles were markedly dilated, and signs of obstructive hydrocephalus with aqueductal stenosis were present (Fig. 1). The enhanced CT scan showed a huge mass, with relatively homogeneous contrast enhancement, in the median part of the posterior fossa (Fig. 2a). The sagittal enhanced reconstruction CT showed a high density mass extending from the upper border of the fourth ventricle roof to the incisura and forward to the quadrigeminal cistern (Fig. 2b). In the arterial phase of vertebral arteriography, the vermian and hemispheric branches of the posterior inferior cerebellar artery (PICA) were stretched and displaced outward so as to surround the tumor. The basilar artery was displaced forward and stretched, and the superior cerebellar artery displaced upward, giving the appearance of a tight posterior fossa. No tumor stain was seen in the arterial phase (Fig. 3 a, b). In the venous phase, a faint tumor stain was noted in the median part of the posterior fossa; in the lateral part, an image of irregular venous congestion was noted (Fig. 3 c).

A V-P shunt was first inserted for the internal hydrocephalus. One month later, removal of the tumor was attempted. The patient was placed in a prone position, and reconstructive suboccipital craniotomy was performed with a median incision in the occipitonuchal region. The dura mater was incised in the shape of a Y. With the incision of the dura a reddish purple smooth-surfaced tumor protruded (Fig. 4a). The tumor was situated in the cerebellar vermis, with a part embedded in a saw-tooth manner in both cerebellar hemispheres. On detailed observation, thin folia of the cerebellar hemispheres were found to cover the surface of the tumor (Fig. 4 b). Vaporization with a CO_2 laser was started from the surface of the tumor and excision en bloc attempted, but profuse bleeding was encountered. Consequently, bipolar coagulation was used while compressing the site of bleeding, and the tumor was excised as rapidly as possible. When some space had been obtained, the entrance of the vermian branches of PICA into the tumor were sectioned by coagulation. Bleeding rapidly decreased. The CO_2 laser was then used to remove the tumor en bloc. Next, the residual tumor attached to the paracollicular region was removed under a surgical microscope. After total removal, the vein of Galen, the dorsal part of midbrain and floor of the fourth ventricle, as well as the caudal part of medulla,

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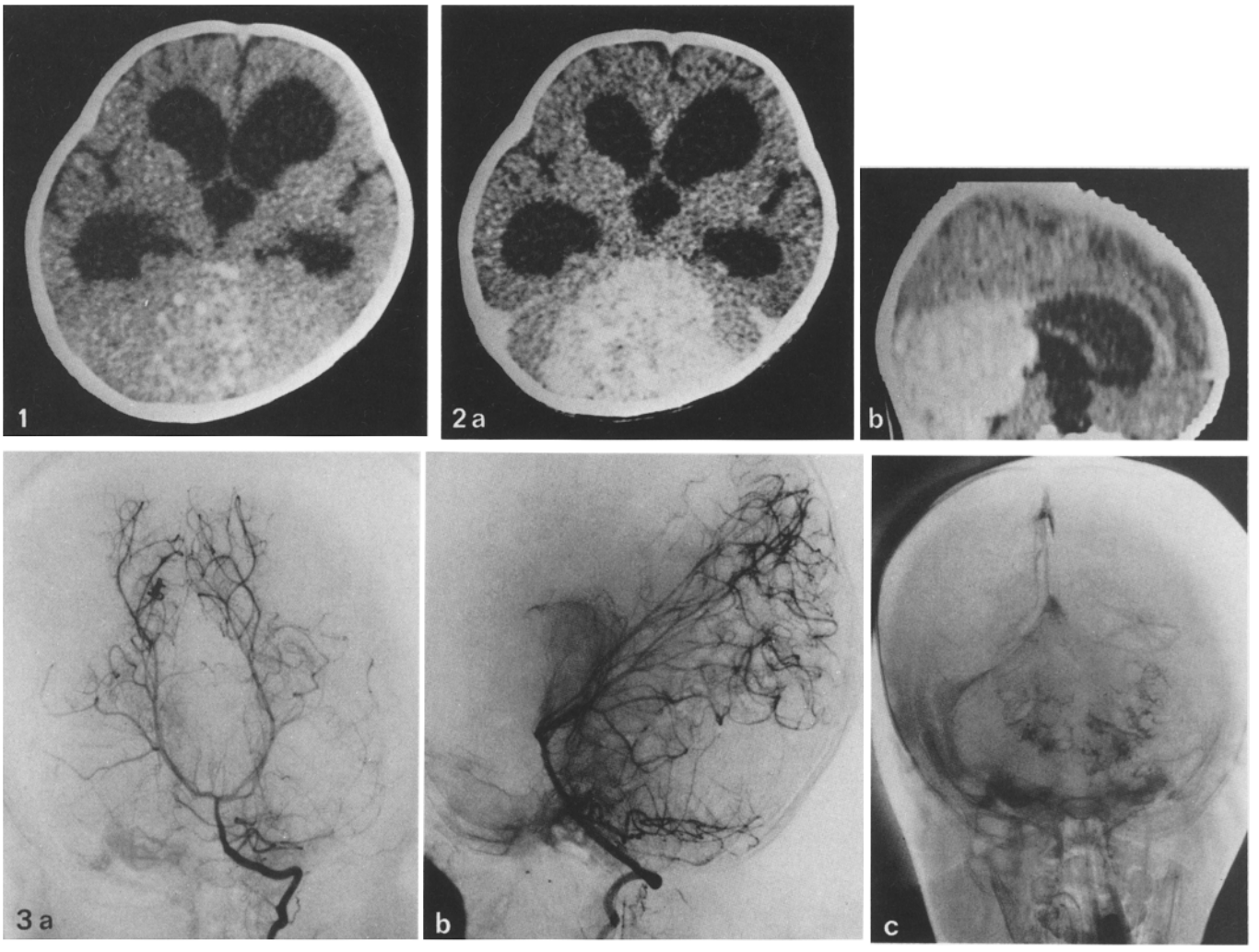


Fig. 1. Plain CT shows spotty, high density zone in the median part of the posterior fossa and markedly dilated lateral and third ventricles

Fig. 2a, b. Enhanced CT shows a homogeneous contrast enhancement in and around the cerebellar vermis (**a**), and the midsagittal enhanced reconstruction CT shows an enhanced mass in the shape of enlarged cerebellar vermis, and dilated lateral and third ventricles (**b**)

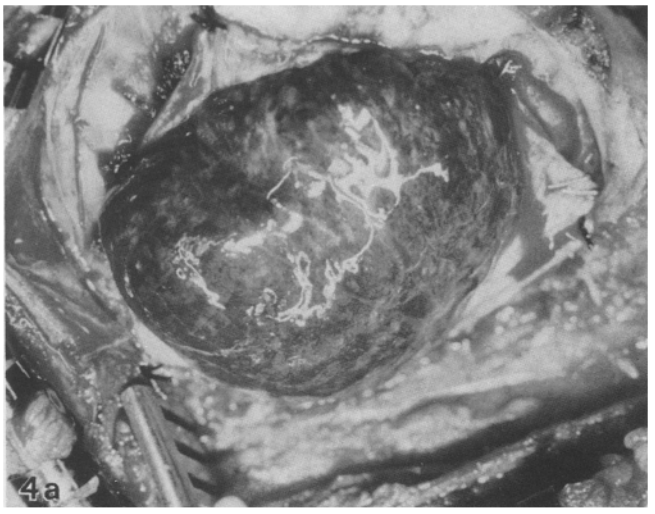
Fig. 3a–c. In the arterial phase of the left vertebral arteriography, the vermian and hemispheric branches of PICA are stretched and displaced outward so as to surround the tumor. The basilar artery is displaced forward and stretched, and the superior cerebellar artery is displaced upward (**a, b**). In the venous phase, a faint stain is noted in the tumor site with engorged, tangled, efferent veins in the marginal part irregular (**c**)

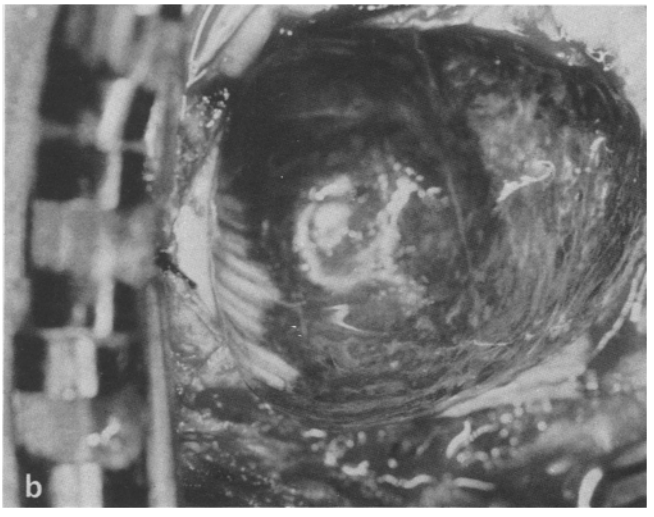
were exposed. In the first postoperative week there was disturbance of consciousness and respiration, and control by artificial respiration was required. There was a gradual recovery and 2 months later, the patient was discharged from the hospital. In the postoperative CT, a low density area remained at the site corresponding to the tumor. About 1 year after the operation at the age of 1.5 years coordination of all limbs became good and standing became possible when holding onto an object (Fig. 5).

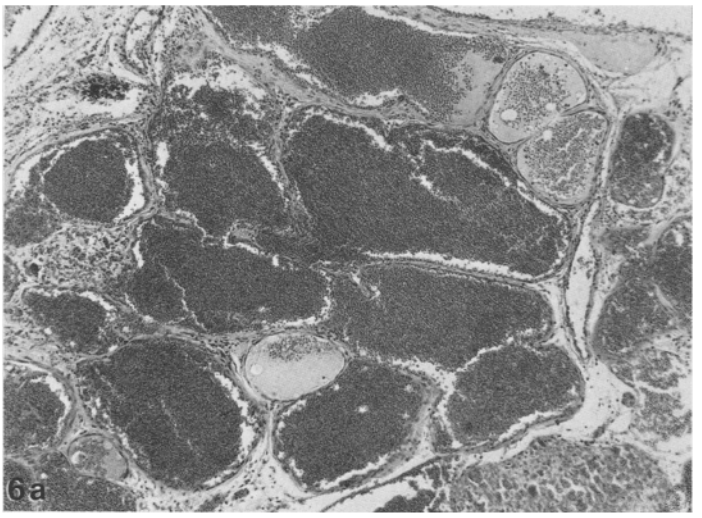
Histology of the tumor revealed an aggregation of dilated blood vessels covered with a layer of endothelial cells, giving the appearance of a cavernous hemangioma in some parts (Fig. 6a). This extended over a wide area of cerebellar tissue, containing granular cells and a few Purkinje cells, intermeshed irregularly amidst vascular channels (Fig. 6b). Blood vessels in the angiomatous lesion showed a layer of endothelial cells without mitosis and a surrounding collagenous layer, but no smooth muscle. This angioma can, therefore, be interpreted as a vascular hamartoma in which cavernous hamartoma and telangiectasia are mixed. The cerebellar tissue in the angiomatous lesion showed an outer granular layer in some parts, but in most sites the structure of the folia was indistinct. From these findings, the lesion can also be considered to be a cerebellar anomaly associated with angioma. Presence of cavitation or blood pigment indicating previous hemorrhage was not observed.

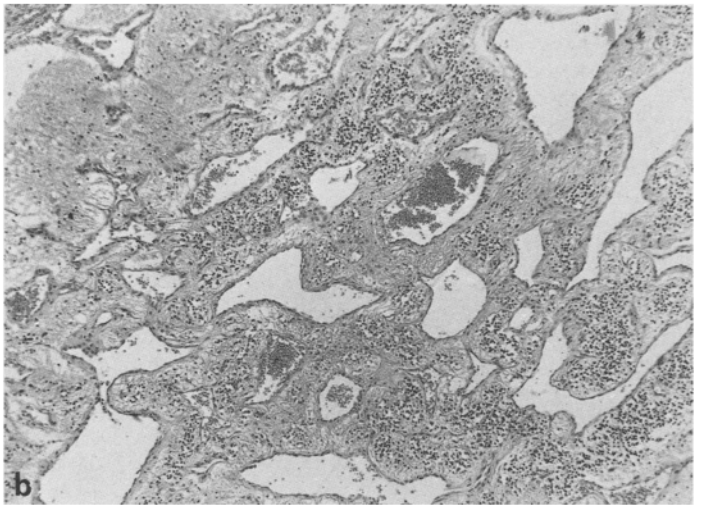
Discussion

Various classifications of angioma of the brain have been proposed by Virchow [15], Cushing and Bailey [3], Bergstrand [2], and Russell and Rubinstein [13]. The last of









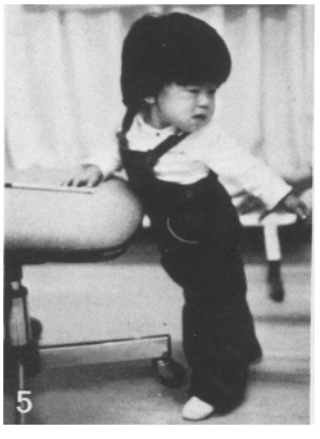


Fig. 4a, b. Intraoperative photographs show a protruded, reddish-purple smooth-surfaced tumor (**a**) and the folia of the cerebellar hemisphere extending over the surface of the tumor (**b**)

Fig. 5. Photograph of the patient 1 year after operation

Fig. 6a, b. The excised specimen shows aggregated, dilated blood vessels covered with a layer of endothelial cells, giving the appearance of a cavernous hemangioma (**a**), and dilated vascular channels intervened by the cerebellar tissue, containing granular cells and a few Purkinje cells, giving the appearance of telangiectasia (**b**). ×93, H&E stain

these is the most widely used today. According to this classification, cavernous angioma, capillary telangiectasia, and venous or arteriovenous malformations are all considered vascular hamartomas, having no characteristics of a true neoplasm, and interpreted as a congenital vascular anomaly. The most frequent site for an angioma in the brain is the cortex in the Rolandic area, the subcortical area, and the basal ganglia and pons [13]. Generally, the rate of occurrence in the cerebral hemispheres is said to be as high as 70%–90%, while the incidence in the posterior fossa, including the brain stem, is said to be 5%–17% [1, 6, 16].

Other than in arteriovenous malformation, angioma rarely occurs in the cerebellum. Only 4 cases were found among 510 cases reported by McCormick [8] and 4 among 164 cases of intracranial cavernous angioma compiled by Voigt and Yasargil [16]. As far as we know, no report

exists regarding a huge cerebellar angioma as seen in our case. In addition, the age of onset of symptoms of angioma in the brain is mostly in the 20–50 year range: onset after 30 years of age is seen in most cases [14]. An onset under 1 year of age is very rare. Intracranial cavernous angioma in the newborn was found in the choroid plexus of the lateral ventricle by McGuire et al. [9] and Iwasa et al. [7], and in both cases the chief sign was hydrocephalus. These are the only reports we have found.

Our angioma showed features of cavernous hemangioma in some parts, but contained blood vessels, which can be considered telangiectasia over a wide area: a combination of cavernous hemangioma and telangiectasia. Such cases of combined vascular anomalies have been discussed at length by Russell and Rubinstein [13]. Further, in our case, formation of folia by cerebellar tissue is poor at the site where angioma is present. Consequently, it can also be interpreted as a cerebellar anomaly which contained an angioma.This corresponds well with the fact that the enhanced mass in the midsagittal reconstruction of the CT scan showed the morphology of an enlarged vermis.

The radiological characteristics of vascular anomaly in the brain show a wide range of variation, so that correct preoperative diagnosis is often difficult. Cavernous hemangioma often appears as a high density mass in the CT scan, showing marked contrast enhancement, but a case without enhancement has also been reported [5]. Angiography usually shows an avascular mass; however, a vascular stain in the venous phase [10] or a highly intense stain [5] has also been reported. Telangiectasia is often difficult to observe by angiography, but it is reported that a blush may appear in the intermediate phase of angiography and dilated channels may be seen in the venous phases [10]. There are few reports on the combination of cavernous hemangioma and telangiectasia, but Robertson [12] has reported a case showing early filling of veins and dilated efferent veins around an avascular mass, which are somewhat similar to the findings in our case.

The usual angioma seldom shows a marked mass effect, unless bleeding occurs. However, in our case, this was seen at the time of surgery, even after the insertion of a V-P shunt for hydrocephalus. In the tumor, no hematoma was present and the mass effect was due to the huge bulk of the hemangioma itself, though it was histologically benign. The only treatment was removal of the tumor in our case. This resulted in the whole vermis and the median portions of both cerebellar hemispheres being lost after tumor removal. However, the patient was able to stand up while holding on to an object 1 year after surgery. Because of the fact that coordination and equilibrium are developing, though delayed, it is assumed that the function of the cerebellar vermis is compensated by the residual cerebellum. Such a plastic recovery is likely to occur more prominently when surgery is undertaken in infancy than in older childhood or in adults.

References

1. Bebin J, Smith EE (1982) Vascular malformations of the brain. Raven Press, New York, pp 13–29 (Seminars in neurological surgery)
2. Bergstrand H, Olivecrona H, Toennis W (1936) Gefäßmißbildungen und Gefäßgeschwülste des Gehirns. Thieme, Leipzig
3. Cushing H, Bailey P (1928) Tumors arising from the blood vessels of the brain. Thomas, Springfield, Ill
4. de Tribolet N, Kaech D, Perentes E (1982) Cerebellar haematoma due to a cavernous angioma in a child. Acta Neurochir 60:37–43
5. Fukui M, Matsuoka S, Hasuo K, Numaguchi Y, Kitamura K (1983) Cavernous hemangioma in the pineal region. Surg Neurol 20:209–215
6. Giombini S, Morello G (1978) Cavernous angiomas of the brain, account of fourteen personal cases and review of the literature. Acta Neurochir 40:61–82
7. Iwasa H, Indei I, Sato F (1983) Intraventricular cavernous hemangioma. J Neurosurg 59:153–157
8. McCormic WF, Hardman JM, Boulter TR (1968) Vascular malformations (angiomas) of the brain, with special reference to those occurring in the posterior fossa. J Neurosurg 38:241–251
9. McGuire TH, Greenwood J, Newton BL (1954) Bilateral angioma of choroid plexus. Case report. J Neurosurg 11: 428–430
10. Numaguchi Y, Kishikawa T, Fukui M, Kitamura K (1979) Prolonged injection angiography for diagnosing intracranial cavernous hemangiomas. Radiology 131:137–138
11. Pozzati E, Padovani R, Morrone B, Finizio F, Gaist G (1980) Cerebral cavernous angioma in children. J Neurosurg 53: 826–832
12. Robertson GH, Kase CS, Wolpow ER (1974) Telangiectasis and cavernous angiomas of the brain stem: "cryptic" vascular malformations. Report of a case. Neuroradiology 8:83–89
13. Russell DS, Rubinstein LJ (1971) Pathology of tumours of the nervous system, 3rd edn. Williams & Wilkins, Baltimore, pp 93–98
14. Savoiardo M, Strada L, Passerini A (1983) Intracranial cavernous hemangiomas: neuroradiologic review of 36 operated cases. AJNR 4:945–950
15. Virchow R (1982) Vascular malformations of the brain. Raven Press, New York, pp 13–29 (Seminars in neurological surgery)
16. Voigt K, Yasargil MG (1976) Cerebral cavernous haemangiomas or cavernomas. Incidence, pathology, localization, diagnosis, clinical features and treatment. Review of the literature and report of an unusual case. Neurochirurgia 19:59–68