Cavernous hemangioma extending to extracranial, intracranial, and orbital regions

Case report

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Extraaxial cavernous hemangiomas are rare intracranial lesions that can be located in different cranial compartments. Extension across different tissue planes such as the subcutaneous tissue, skull, orbital cavity, intracranial dura mater, and extracranial trigeminal divisions within the same patient has not been previously reported.

This 32-year-old woman suffered left exophthalmos, left sixth nerve palsy, and trigeminal neuropathy. Magnetic resonance imaging studies revealed an extensive multicompartamental lesion, with enhancement following Gd administration.

A left orbitopterional approach allowed removal of several cavernomatous lesions located in the orbit, frontotemporal dura, and lateral wall of the cavernous sinus. A histologically based diagnosis of extraaxial cavernous hemangioma was made. In the postoperative period the patient experienced a regression of her symptoms.

The authors report on a case of cavernous hemangioma with a unique extension to different intracranial/extracranial compartments. Although radical removal of the lesion was not feasible, partial excision allowed for satisfactory clinical control of the patient’s symptoms.

KEY WORDS • cavernous hemangioma • vascular malformation • orbital tumor • magnetic resonance imaging
Biopsy. Biopsy specimens of a small (2 cm) subcutaneous left retromastoid lesion were obtained after the administration of a local anesthetic agent. These tissue specimens were soft and had a dark reddish spongy appearance. After fixation in 10% buffered formalin, the samples were embedded in paraffin using standard procedures. Tissue sections (5 μm thick) were stained with H & E. Through a microscope, the tissue appeared quite uniform, consisting of large collections of vessels with large lumens filled by numerous red blood cells. The endothelium was minimally apparent and the wall thin and fibrous. Some areas were hemorrhagic with the presence of red blood cells free in the intervascular space. The tumor appeared to be completely benign and the diagnosis of cavernous hemangioma was made.

Second Admission. Three years after the biopsy procedure, the patient experienced continuous paresthesia in the left fifth cranial nerve distribution. Clinical examination showed mild progression of the left exophthalmos and moderate hypesthesia in the first two divisions of the left trigeminal nerve. An MR image and a cerebral angiogram confirmed the aforementioned findings and revealed a slight increase in the size of both the intraorbital and intracranial mass lesions. Surgical decompression of the orbit and the cavernous sinus was proposed but not accepted by the patient.

Third Admission. At the last admission, 6 years after the onset of symptoms, a progressive increase in the left exophthalmos and a sixth nerve palsy were evident. Magnetic resonance images (Fig. 1A–F) demonstrated the presence of the multiple left intraorbital and intracranial/extracranial mass lesions, with dural nodules in the posterior fossa and supratentorial frontotemporal areas. The most impressive features of the lesions included a high signal intensity on T2-weighted images, a low signal intensity on T1-weighted images, bright contrast enhancement after contrast injection, appearance of multiple adjacent round components, a tendency to grow along the left trigeminal nerve roots (especially along the mandibular division), enlargement of the left superior orbital fissure, and the coexistence of other ipsilateral subcutaneous locations and contralateral right foramen ovale involvement. Magnetic resonance angiography revealed no sign of high-flow vascular malformations.

The patient underwent a left orbitopterional craniotomy for the removal of several intraorbital soft encapsulated masses as well as three small cavernomas located in the frontotemporal dura (Fig. 2). En bloc removal of each le-
Extracranial, intracranial, and orbital cavernous hemangiomas

Fig. 2. Intraoperative photographs showing intraorbital (A) and intradural frontotemporal (B) extraaxial cavernomatous lesions.

Clinical and neuroradiological discrepancies between intra- and extraxial cavernomas are explained by their different locations. These lesions are bone erosion or scalloping, isodensity or hyperdensity, and can be uni- or multifocal. Skull base involvement has been reported, whereas inner table and extradural space are rarely affected. Recently, a very large cavernoma of the convexity involving several adjacent tissues (subcutaneous, skull, and extradural space) was described. Note also that an ECH can extend to the subarachnoid space and cause subarachnoid hemorrhage.

Cavernous angiomias of the orbit are the most frequently occurring benign lesions in this location and must be differentiated from orbital lymphangiomas. The most common clinical symptoms of cranial ECHs are headache, cranial neuropathy, exophthalmos, and vision loss. Pertinent radiological studies include computerized tomography scanning, angiography, and MR imaging. Common features on computerized tomography scanning are bone erosion or scalloping, isodensity or hyperdensity, and homogeneous enhancement after iodinated contrast injection; the demonstration of a dual tail is very unusual.

Discussion

Extraaxial cavernous hemangiomas are rare intracranial vascular lesions that have been the subject of a single case or a small series. They have been more frequently reported on by Japanese authors and are associated with a female predominance in most series. These hemangiomas show a tendency to grow, which could be due to neoplastic progression; peripheral capillary outgrowth, ectasia of vascular spaces, and thrombosis with progressive sclerosis have also been proposed as possible explanations. Interestingly, some authors consider ECHs to be proliferative neoplastic lesions, similar to hemangiomas occurring in infancy. Nevertheless, this concept is not accepted by others who define these lesions as vascular congenital malformations very similar histologically and immunohistochemically to parenchymal cavernomas.

Postoperative Course. The postoperative course was uneventful. At the first clinical follow up 2 months after surgery, the patient was in good clinical condition, showing regression of the left exophthalmos and the sixth nerve palsy. Thirteen months later the patient was again admitted to our institution for the removal of a painful subcutaneous left parietal hemangioma; the lesion had increased in size during the past year, reaching a diameter of 3 cm. Neurological examination confirmed the stability of the moderate left exophthalmos and the mild left trigeminal hypesthesia. Visual acuity and ocular motility were normal. An MR image demonstrated satisfactory orbital and anterior skull base decompression with no sign of cavernomatous regrowth in these regions. No adjuvant therapy was proposed during the follow-up period, either for the regression of the patient’s main complaints or for the risk of radiotherapy.

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Extracranial, intracranial, and orbital cavernous hemangiomas

Fig. 3. Photomicrograph with low-power magnification showing large dilated vessels lined by flattened endothelium. H & E, × 25.
Small calcifications may be found as in other angiomas. On MR imaging, ECHs are usually isointense or hypointense on T₁-weighted images, brightly hyperintense on T₂-weighted images, and intensely contrast enhancing. Angiographic features include vessel dislocations and necrotic tumor blush. Differential diagnosis includes meningiomas, neuromas, and high-flow vascular malformations (fistulas and arteriovenous angiomas). A correct preoperative diagnosis could be very important in planning the surgical approach, but in most cases a definitive diagnosis can be reached only on histological study. The most significant MR imaging findings for a correct differential diagnosis appeared to be the bright hyperintensity on T₁-weighted images, marked contrast enhancement, and absence of infiltrative bone involvement despite extensive growth in the cranial base. Skull base meningiomas can demonstrate peculiar angiography findings of vessels radiating outward from the central pedicle as well as MR imaging findings of signals similar to gray matter on both T₁-weighted and T₂-weighted images, dural tail, evidence of a smooth angle between tumor and dura mater, and calcifications. Surgical removal could be difficult because of the vascularity and related intraoperative blood loss as well as cranial nerve involvement and internal carotid artery encasement. Total removal has been reported in only 30 to 35% of the cases. Morbidity and mortality rates have been high. Because of the surgical difficulties, radiotherapy has been proposed as preoperative or complementary treatment or as primary therapy in debilitated patients. Proposed surgical biopsy followed by radiotherapy before undertaking surgery, but the radiosensitivity of cavernous hemangiomas is unpredictable. Preoperative embolization can be useful in highly vascularized lesions.

Conclusions

The singular feature in the patient in this report is the hemangioma location in different compartments such as soft tissue, skull, orbital cavity, cranial base dura, and temporalis as well as the extracranial spreading along the third division of the fifth nerve. A slowly progressive clinical course is typical of a benign neoplastic lesion instead of a true vascular malformation. The widespread extension precluded any form of radical treatment; however, surgical removal of the orbital and frontotemporal lesions was followed by regression of the patient's main disturbances without any new postoperative deficit.

References


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