Dural Cavernous Hemangioma: An Under-recognized Lesion Mimicking Meningioma

James R. Perry, William S. Tucker, Mario Chui and Juan M. Bilbao

ABSTRACT: We report a 77-year-old woman who presented with partial seizures and was found to have an enhancing dural-based parietal convexity mass. The lesion enlarged on serial examination by computed tomography (CT) over a one year period. The clinical features and radiologic appearance were compatible with a pre-operative diagnosis of meningioma; however, pathologic findings were typical of a dural cavernous hemangioma. Accumulating evidence suggests that these lesions are an uncommon but distinct type of vascular malformation most often arising from the cavernous sinus, tentorium, or cerebello-pontine angle. With CT, magnetic resonance imaging and angiography, these lesions can closely resemble meningioma in terms of signal characteristics, enhancement pattern, and location. This is of importance both in the practical management of meningiomas where the diagnosis is often based on radiologic studies alone, and in clinical trials where incorrect entry diagnosis should be avoided.

RÉSUMÉ: Malformation caverneuse d'origine durale: une entité peu reconnue. Nous rapportons le cas d'une femme de 77 ans qui a présenté des crises d'épilepsie partielle, chez qui on a découvért une masse rehaussante d'origine durale à la convexité pariétale. La lésion a augmenté de volume sur une période d'un an, tel que démontré par des examens sériés par CT scan. Les manifestations cliniques et les images radiologiques étaient compatibles avec le diagnostic pré-opératoire de méningiome; cependant, l'anatomopathologie fut typique d'un hémangiome caverneux dural. Nous avons de plus en plus d'évidence que ces lésions constituent un type peu fréquent mais distinct de malformation vasculaire, prenant naissance le plus souvent à partir du sinus caverneux, de la tente du cervelet ou de l'angle ponto-cérébelleux. Au CT scan, à la résonance magnétique et à l'angiographie, ces lésions peuvent ressembler au méningiome quant aux caractéristiques du signal, à l'aspect du rehaussement et à la localisation. Ceci est important, tant pour le conduite du traitement des méningiomes, où le diagnostic est souvent basé seulement sur les études radiologiques, que pour les essais thérapeutiques où on doit éviter d'inclure un individu dont le diagnostic est erroné.

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Cavernous hemangiomas (cavernomas) comprise 5-13% of central nervous system vascular malformations.[1,2,3] The majority are intra-axial occupying the cerebral hemispheres or, occasionally, the cerebellum or brainstem. Extra-axial cavernous hemangiomas arise in relation to dura in the middle fossa, cerebello-pontine (CP) angle, and along the tentorium but represent only a small fraction of all such lesions.[3] The clinical and radiologic features of dural cavernous hemangiomas have not been well documented; but, unlike the classic parenchymal lesions, they have been noted to resemble meningioma in past reports.[3]

The computed tomography (CT) appearance of meningioma is highly characteristic. Atypical features such as irregular enhancement, low density areas, and edema are so frequent that an accurate pre-operative diagnosis can still be made.[4,5,6] In a National Cancer Institute sponsored study, enhanced CT accurately detected 96.2% of intracranial meningiomas.[4] Decisions on patient management are thus often based on a clinical and radiologic diagnosis alone. Elderly patients may be treated conservatively and more recently, trials of therapy such as hormonal manipulation[7] and radio-surgery[8] are being conducted without histologic diagnosis in some cases. Accuracy of diagnosis has therefore become important both on practical clinical grounds and for the purposes of research trials.

We describe the case of a dural cavernous hemangioma arising from the parietal convexity of a 77-year-old woman. This is, to our knowledge, the first description of a cavernous hemangioma in this location. The clinical and radiologic findings over an 18 month period were consistent with meningioma and led to misdiagnosis pre-operatively.

Case Report

A 77-year-old woman in excellent physical health presented with partial seizures involving her right arm. Her neurologic examination was normal. A CT scan showed an irregularly enhancing, 3.5 cm diameter, extra-axial mass lesion arising from the left parietal convexity (Figure 1B). Her seizures were well controlled with anticonvulsants and she elected to be followed conservatively. One year later she developed recurrent seizures and progressive right hand weakness. Repeated CT demonstrated slight enlargement of the lesion but no further change. A diagnosis of meningioma was made.

The patient underwent left parietal craniotomy. Dura was adherent to overlying bone that was normal in appearance. A dark,vascular mass

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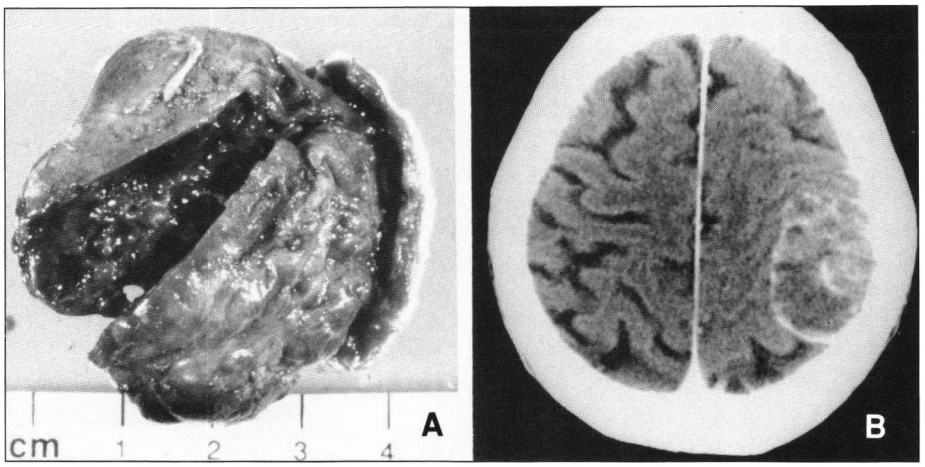

Figure 1A — Operative specimen demonstrating a vascular mass attached to dura. The cut surface consists of large dilated channels that, microscopically, are lined by a single layer of endothelium.

1B — Enhanced CT scan at the time of presentation with focal seizures. A well-demarcated, enhancing convexity mass is noted. No bony change was evident.

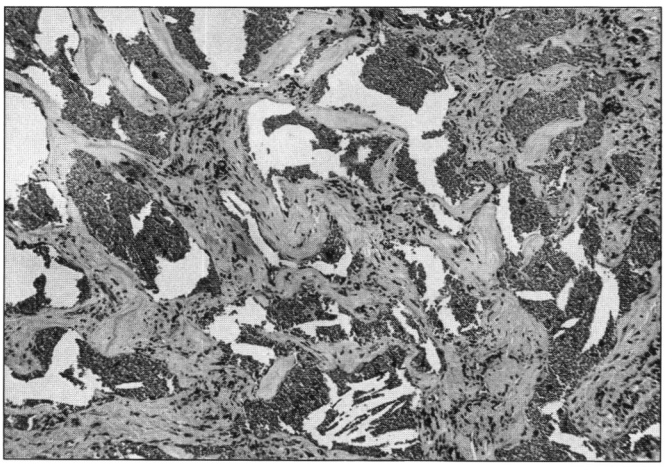

Figure 2 — Representative area of the lesion showing prominent and closely packed veins with hyalinized and fibrotic walls (hematoxylin and eosin, original magnification X 80).

was encountered upon opening the dura. It mobilized easily and was dissected away from brain en bloc (Figure 1A). Cortex below the lesion appeared effaced but normal. Apart from transient seizures immediately post-operatively, she made an uneventful recovery.

Pathologic examination revealed a soft, dark red vascular mass attached to dura. The cut surface revealed numerous large dilated vascular channels and cystic spaces. No portion of the lesion was solid or nodular. Histologically, the channels were lined by a single layer of endothelium (Figure 2). No intervening neuropil or elastic containing vessels were present. Features of old hemorrhage with subsequent organization including hemosiderin deposition, cholesterol clefts, focal calcification and fibrosis were prominent. These findings are characteristic of cavernous hemangioma.[9]

Discussion

Meningiomas are the most common extra-axial mass lesions in adults.[9] Clinical decisions are often made based upon their characteristic appearance on widely available CT imaging. Although the diagnostic accuracy of CT reached 96% in a large clinical study, the false positive rate was 1.9 percent.[4] In that study, 50 of 2,555 patients with intracranial tumours were given an incorrect diagnosis of meningioma on radiologic grounds. The final diagnoses in these cases were glioma, normal, metastasis, schwannoma, other tumours, non-neoplastic lesions, and cerebral infarction in descending order of frequency. In many reports, the opposite situation has been noted; that is, lesions diagnosed as other neo plasms radiologically were found to be meningioma post-operatively.[10] This has led to a wide literature on the "atypical" CT features of meningioma including cyst formation, irregular enhancement, low density regions, peritumoural edema, and hemorrhage.[6,11] In our case, the CT appearance included the typical features of extra-axial location, a broad dural base, and sharp demarcation from underlying brain. "Atypical" features included irregular enhancement, and an area of hypodensity within the lesion. Focal calcification was seen pathologically but was not seen on plain CT imaging.

Location

Cavernous hemangiomas are thought to uncommonly arise in relation to dura; however, recognition of their full spectrum of expression is increasing, particularly since the advent of magnetic resonance imaging (MRI).[1,3] In a review of 138 histologically-confirmed cavernous hemangiomas, 13 were found extra-axially in the middle fossa, four in the CP angle, and one associated with the tentorium. In some of these cases both the clinical syndrome and CT findings suggested a diagnosis of meningioma.[3] Since the time of that review in 1986, extra-axial cavernous hemangiomas have been reported in the cavernous sinus,[12-15] the CP angle,[16] tentorium,[17] Meckel's cave,[18] cauda equina,[19] cranial nerves,[20] anterior cranial fossa,[21] and as a congenital convexity lesion in a neonate.[22] The overall distribution is therefore remarkably similar to meningiomas in general,[23] with over-representation of the cavernous sinus lesions, possibly suggesting a predisposition for that site.[13,24] Other similar clinical features include both female preponderance and enlargement during pregnancy.[13,21]

Radiologic Appearance

The CT appearance of dural cavernous hemangiomas, like meningioma, is non-specific and shows a well-defined extra-axial mass lesion on a broad dural base, with regular contrast enhancement. Plain skull radiographs or bone windows with CT are normal or reveal bony erosion involving the dorsum sella, posterior clinoid, or floor of the middle fossa.[13,24] Calcification is uncommon radiologically, and well-defined bony hyperostosis has not been reported. These latter features allow some distinction from meningioma; however, calcification is seen in only

11% and hyperostosis in only 15% of proven meningiomas on plain radiographs, with CT adding minimally improved sensitivity.[4]

The MRI characteristics of dural cavernous hemangiomas have not been well described. Clearly MRI has added increased sensitivity in the detection of parenchymal lesions and is the imaging tool of choice for these.[2] In the cavernous sinus, well demarcated dural-based enhancing lesions on MRI have been reported.[12-15] Unlike typical intra-axial cavernous hemangiomas with mixed signal intensities centrally and a rim of signal void,[1] the extra-axial lesions have shown homogeneous signal characteristics, are isointense or hyperintense on T2 sequences, and enhance strongly with gadolinium.[12-15] These features closely resemble those of meningioma and are inseparable in many cases.[12,13]

Cerebral angiography has been performed in previous cases, revealing no abnormality in one-third, or an avascular mass with some displacement of adjacent vessels in two-thirds.[21] Less commonly, an angiographic tumour blush is noted especially with middle fossa lesions.[13,24] Angiographic findings can therefore be non-specific, and as in our case, angiography may not be performed when CT or MRI findings are felt to be diagnostic and the lesion is surgically accessible.

Management

Management has been primarily surgical, especially with easily accessible locations such as in our case. With deeper lesions the role of radiation, radiosurgery, or combined treatment with surgery is unclear. Cavernous sinus lesions have been reported responding to external radiotherapy[14,25] and while intra-axial cavernous hemangiomas may be at least partially radiosensitive, the role of radiosurgery remains unclear.[25,26] Traditionally, these dural lesions are thought to be difficult to excise because of intra-operative bleeding and adherence to adjacent structures such as the cranial nerves.[3,13,21] In our case complete excision was possible with minimal bleeding and this has been the experience in other recent reports.[13,16] Clearly, location plays an important role and MRI should improve operative planning as recognition of these lesions increases.

Pathology and Mechanism of Growth

Histologically, the lesion in our case was typical of a cavernous hemangioma. Of interest, it was seen to enlarge over a relatively short period of time (18 months) causing further clinical concern and confusion with a neoplasm. The precise mechanism of growth in these lesions is unclear but is believed to be related to repeated micro-hemorrhage, calcification, and cholesterol accumulation.[1] These growth features were seen abundantly in our case. Of note, the histologic features of cavernous hemangiomas arising specifically from the dural sinuses has been unique. Many cases have demonstrated only large sinusoidal channels and lack the other features typical of cavernous hemangioma.[12,13] These particular lesions may be separable from other dural cavernous hemangiomas and the term "sinus cavernoma" has been proposed.[13] Growth has been well documented in previous cases of intra-axial cavernomas and their natural history has been well studied.[26] Reports have appeared of cavernous hemangiomas associated with neoplastic glial tissue and there is some evidence that they can be induced by polyoma virus in animal models.[27] This concept has been challenged and these so-called "angiogliomas" likely represent unusually vascular low-grade gliomas rather than a distinct entity.[28]

We believe that dural cavernous hemangiomas are underrecognized. There is increasing evidence that they represent a distinct subgroup of vascular malformations in general,[29] and often mimic meningioma on both clinical and radiologic grounds. Further study is required to identify criteria that may help to distinguish these lesions from other dural-based masses. We recommend that dural cavernous hemangioma be included in the differential diagnosis of meningioma, especially in the presence of "atypical" clinical or radiographic features, and particularly for patients being managed with novel therapy or being enrolled into clinical trials.

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