Hypertrophic cranial pachymeningitis: assessment with CT and MR imaging.

N Martin, C Masson, D Henin, D Mompoint, C Marsault and H Nahum

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Hypertrophic Cranial Pachymeningitis: Assessment with CT and MR Imaging

Three patients with diffuse idiopathic cranial pachymeningitis with predominant involvement of the tentorium and falx are reported. Progressively increasing headaches were the usual symptoms, along with ataxia and various cranial nerve palsies. CT in all cases and MR imaging in two cases detected isolated thickened dura mater. In one case, MR depicted dural involvement as a very large, hypointense area with fine hyperintense edges on T2-weighted images. Microscopic examination of thickened dura revealed extensive fibrotic tissue with a chronic inflammatory infiltrate containing lymphocytes, plasma cells, and scattered eosinophils; these findings closely paralleled the MR features. Only four cases with similar pathologies have been described, all before the advent of CT and MR. We discuss the different causes of thickened dura mater as well as the significance of the fact that dural thickening can be responsible for occlusion of the dural sinuses.

Cranial pachymeningitis is a rarely reported disease that can resemble other disorders associated with tentorial thickening; CT and MR can help differentiate it from these other disorders.

Idiopathic hypertrophic cranial pachymeningitis is a rare disease. Only a few clinical cases were reported before the advent of CT. We describe three cases of isolated dural thickening and discuss possible causes with reference to CT, MR, and clinical and pathologic data.

Materials and Methods

Three patients, two women and one man aged 20, 58, and 58 years, with severe headaches were first evaluated by CT before and after administration of IV contrast material.

In two cases, MR studies were performed with a 0.5-T superconducting magnet.* All images were acquired with a 256 × 256 matrix. T2-weighted sequences were obtained with a spin-echo (SE) technique, 1600/60, 120 (TR/TE), with 7-mm-thick contiguous coronal images. A partial-saturation (PS) sequence, 400/12, was obtained only in one case with contiguous 6-mm-thick sagittal images.

Angiograms were obtained in two cases, and special attention was given to veins and dural sinuses. All patients underwent open biopsy. Surgical and pathologic results were noted.

Case Reports

Case 1

A 20-year-old woman was first seen in January 1986 for evaluation after 6 months of progressively increasing severe left-sided headaches, which at the time of admission occurred daily and sometimes were associated with vomiting. No abnormalities were found at neurologic and general examinations. No papilledema was seen.

Skull radiographs were normal. CT demonstrated extensive changes (Figs. 1A–1E). A technetium brain scan revealed slight left-sided tentorium enhancement. Vertebral and carotid
Fig. 1.—Case 1.
A–E, CT scans show asymmetric, well-defined, irregular, and nodular thickening of tentorium gradually decreasing laterally.
A and B, Axial (A) and coronal (B) precontrast scans show heterogeneously increased attenuation of thickened tentorium, 19 mm wide on left (arrowheads).
C–E, Postcontrast CT scans show marked enhancement prevailing on edges. No extension is seen along falx (D). Note mass effect on cerebellar hemisphere with displacement of V₄ (E). No parenchymal abnormalities are seen.
F and G, Angiograms show narrowing of posterior part of superior sagittal sinus (straight black arrow) with poor filling of right transverse sinus (white arrows); left transverse sinus is invisible (curved arrow).
H, PS image, 400/12, left parasagittal section. Thickened tentorium appears as slightly hypointense area relative to adjacent cerebellar cortex.
I and J, T₂-weighted coronal images show very-well-delineated lesion extending throughout dura, bearing two components.
I, SE 1800/60 image. Large, central, slightly heterogeneous hypointense area prevails on left tentorium; on superior and inferior edges, a fine line of very high intensity is seen (arrowheads).
J, SE 1800/120 image. Peripheral hyperintensity enhances, also visible on right (arrowheads).
angiograms demonstrated abnormalities in the dural venous sinuses but no arterial abnormalities (Figs. 1F and 1G). The chest radiograph was normal. Erythrocyte sedimentation rate was 78 mm/hr. Blood count, serum and urine electrophoresis, and serum angiotensin-converting enzyme level were normal. The VDRL test was negative. Oligoclonal banding was absent. CSF protein was 0.47 g/l (immuno-globulin G/albumin ratio, 31%) with no abnormal cells.

An open biopsy disclosed a firm, grossly thickened tentorial dura mater. The dura mater was thickened and composed of loose collagen tissue with numerous vessels. Cellular infiltrates of lymphocytes and polyclonal plasma cells with rare eosinophilic or neutrophilic polymorphonuclears and histiocytes were present. Numerous lymphoid follicles were seen. Vessels displayed no arteritis. No epithelioid granulomas, giant cells, or necrosis were observed. Stains and cultures for bacteria or fungi were negative.

On readmission in April 1986, examination revealed abnormal gait and ataxia. Clinical improvement was obtained by steroid therapy. However, headaches reappeared whenever treatment was tapered or discontinued. Follow-up scans were unchanged until January 1987, at which time MR was performed (Figs. 1H–1K). Infratentorial nodularity predominated, but small supratentorial irregularities were also seen. Aside from a cerebellar mass effect, no parenchymal abnormality was displayed.

In July 1987, steroid therapy was interrupted owing to undesired side effects. The patient now receives azathioprine, 100 mg daily, and is asymptomatic.

Case 2

A 58-year-old woman had a 1-year history of severe headaches and vomiting. Past history revealed migraines since infancy. On admission, neurologic and general examinations were normal. However, the first CT scan (May 1981) detected hydrocephalus with tentorial hypertrophy and enhancement after IV contrast material. Development of bilateral papilledema required ventricular derivation. This resulted in regression of the papilledema; persistent headaches resolved with steroid therapy. Recurrence of headaches in December 1981 prompted a complete evaluation. The patient was lethargic and had an unsteady gait. CT revealed small-sized ventricles; thick, strongly enhanced tentorium; and hypodensities of the temporopontinecal white matter.

Left tentorial enhancement was seen on a technetium brain scan, but a gallium brain scan was normal. Chest and skull radiographs were unremarkable. Erythrocyte sedimentation rate was 67 mm/hr. Blood count, serum electrophoresis, and serum angiotensin-converting enzyme level were normal. CSF protein, however, was 1600 mg/dl, and 170 cells/ml (60% lymphocytes) were present, without abnormal cultures. Cultures for mycobacteria and VDRL and Treponema pallidum hemagglutination tests were negative.

On open biopsy, the dura mater was markedly thickened on its inner surface by an irregular, firm, yellowish mass. Some adhesion with the underlying arachnoid was seen. On microscopic examination (Fig. 2A), the dura mater was thickened by a loose fibrous tissue with numerous vessels. Some small foci of eosinophilic necrosis were present without caseation or follicular aspects. Scattered dense infiltrates were seen composed mainly of lymphocytes and plasma cells with few epithelioid cells and very rare multinucleated giant cells. The plasma cells were polyclonal. Stains and cultures for bacteria and fungi were negative. No malignant cells were seen. Under steroid therapy, headaches and gait disorders decreased but recurred whenever treatment was tapered. Between 1982 and 1987, progressive neurologic deterioration was observed.

In March 1987, the patient was readmitted with confusion, lethargy, and unsteady gait. A CT examination was performed (Figs. 2B–2I). No arterial abnormalities were detected on cerebral angiograms, but abnormalities in the dural venous sinuses were demonstrated (Figs. 2J and 2K). MR was not performed owing to a ventricular shunt catheter. Bearing in mind the therapeutic procedures used in neurosarcoidosis, local flash radiotherapy of 25 Gy was administered in April 1987, with no result. Currently, the patient continues to receive steroid therapy, now associated with azathioprine. Headaches have decreased but neurologic deterioration seems to continue.

Case 3

In March 1986, a 58-year-old man first noted diminished left-ear hearing acuity with otalgia. Because the symptoms were thought to be related to serous otitis, steroid therapy was instituted, resulting in resolution of the symptoms. Between April and August 1986, the patient complained of progressive headaches and developed facial palsy and dysphonia. Erythrocyte sedimentation rate was 51 mm/hr. CT was considered normal.

In October 1986, under decreased steroid therapy, the patient was admitted with severe headaches, lethargy, and emaciation. On examination left seventh, bilateral eighth, and right 10th and 11th cranial nerve palsies were present with ataxia.

Precontrast CT demonstrated a slight increase in tentorial attenuation, with uniform enhancement on postcontrast scans. The tentorium seemed thickened, but precise appreciation was difficult owing to the absence of coronal sections (Figs. 3A and 3B). Chest and skull radiographs were normal.

Erythrocyte sedimentation rate was 60 mm/hr. The blood count and panel of serum chemistries were normal. The VDRL test was negative. The CSF contained 104 cells with 100% lymphocytes/ml; the protein level was 440 mg/dl (with oligoclonal gamma globulins, 53%) and glucose level was normal. CSF cytology and culture results were negative.

Open biopsy was performed on temporal dura mater. The dura mater was thickened by dense, fibrous, tissue-containing, small eosinophilic necrotic areas. No caseation was seen. On the edge of the dura, a dense infiltrate of lymphocytes and plasma cells was present with numerous vessels (Fig. 3C). No arteritis was seen. Stains and cultures for bacteria and fungi were negative. No malignant cells were seen. Radiation (30 Gy) was administered and steroid therapy was pursued.

Neurologic examination showed much improvement and steroid therapy was stopped in February 1987. On MR (Fig. 3D), the tentorium was only slightly thickened with a discrete nodular area and a uniform hypointense signal on the T2-weighted sequence. Recurrence of neurologic symptoms led to steroid and azathioprine therapy. In July 1987, CT showed improvement with diminished tentorial enhancement (Fig. 3E), but ventricular dilatation was seen. The patient continues to receive steroid and azathioprine therapy.

Discussion

Normal dura mater is an inert fibrous membrane vascularized by an arterial network in its outer layer and a rich capillary network in its inner layer. On postcontrast CT, tentorial enhancement is always recognizable. Hence, abnormal findings become significant only when the tentorium is thickened or irregular in configuration [1, 2]. Irregular thickening of the tentorium with variable extension to the neighboring dura mater was the common radiologic feature in our three cases, morphologically correlated to a fibrous tissue with lymphocytes and plasma cell infiltrates.
Fig. 2.—Case 2.
A, Microscopic pathology shows inflammatory polymorphic infiltrate with numerous histiocytes, plasma cells, and necrotic eosinophilic areas with pyknotic nuclei. (H and E, X700)
B, Precontrast CT scan shows hyperdense tentorium and posterior falx. Note white-matter hypodensities on left.
C-G, Postcontrast CT scans show marked enhancement and thickening of tentorium and posterior falx. Tentorium is 12 mm thick (E). Posterior falx is 6 mm thick (F, arrows). No enhancement of white-matter hypodensities is seen (G). Ventricles are of normal size.
H and I, Postcontrast CT scans show thickening of posterior part of falx with "empty triangle" sign (arrows).
J and K, Cerebral angiograms.
J, Posterior part of superior sagittal sinus is occluded, with narrowing of straight sinus.
K, Note also narrowing of initial portion of right transverse sinus. Left transverse sinus is not opacified.

**Idiopathic Hypertrophic Cranial Pachymeningitis**

A review of the literature revealed four cases in three reports with clinical and morphologic findings similar to those in our patients [3-5]. However, all were prior to the advent of CT and MR imaging. Some of the cases were originally linked with syphilis, but no definite proof was found and syphilis tests were negative [3, 4].

Severe headaches were constant, sometimes with vomiting, and variously associated with ataxia and cranial nerve
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Fig. 3.—Case 3.
A and B, Postcontrast CT scans show thickening and enhancement of tentorium.
C, Inflammatory infiltrate with prominent infiltration by lymphocytes and plasma cells of internal layer of dura mater, which is thickened by collagen fibrosis. (H and E, x700)
D, SE 1800/60 image shows hypointensity and slightly nodular thickening of tentorium.
E, Section identical to B shows decreased thickening and enhancement of tentorium.

Palsies (VII, VIII, and X), as in our cases. Blindness by optic nerve compression was observed in two cases [3, 5], but was not seen in our patients. The erythrocyte sedimentation rate was always increased. Syphilis tests and germ cultures were negative. Pathologic examination revealed diffuse yellowish thickening of the cerebral dura mater. In the two cases of Michel et al. [3], the thickening prevailed over the tentorium and falx (autopsies) or over the pontocerebellar angle (biopsy) reaching up to 1 cm in thickness, as in the case of Hassin and Zeitlin [4], in which the tentorium was 8 mm thick with extension over the cerebellum and parietal and occipital dura. In the case of Feringa and Weatherbee [5], the skull-base dura mater was 3 mm thick, and over the convexity it was 1 or 2 mm thick, with associated localized bone destruction in the area of the jugular foramen.

Microscopically, the thickened dura mater consisted of considerable fibrosis with infiltrates of chronic inflammatory cells including lymphocytes, plasma cells, and scattered eosinophils. A central, more fibrous area with more inflammatory edges was reported in one of the cases of Michel et al. [3] and was identical to the MR findings in our case 1 and microscopic findings in case 3. Some degree of arachnoid or pia involvement was noted in two cases, but no microscopic brain infiltration was found [3, 4].
Diseases Associated with Tentorial Thickening

Such diffuse dural involvement could not clearly be attributed to a known process, and other diagnostic possibilities may be excluded on the basis of their clinical, microscopic, CT, or MR findings. Most often, the lesion is focal, like meningioma, and dural-based with an eccentric extension. Extensive meningioma-en-plaque or some rare extension of meningiomas throughout the dura can mimic our cases on CT, but extension is more limited and angiograms usually reveal a tumor fed by meningeal arteries. Also, MR imaging displays a marked variability in the signal intensity on T2-weighted images, but never such a hypointense signal as was seen in our patients (Figs. 1H and 1J). The microscopic studies were negative for neoplasm.

Primary intracranial plasmacytoma, arising most frequently from the dura mater of the convexity, has also been reported with a CT appearance similar to that of meningioma.

More diffuse infiltration of the tentorium can be observed in some diseases. Sarcoidosis is a granulomatous disease characterized pathologically by the presence of noncaseating epithelioid granulomas. Up to 5% of cases of sarcoidosis involve the CNS, but intracranial sarcoidosis may be the initial manifestation [6]. Neurologic manifestations may be quite similar to our patients’ symptoms (usually with cranial nerve palsy, aseptic meningitis, etc.) [7, 8]. CT, basal meningitis with hydrocephalus is the most common pattern, variously associated with tumorlike parenchymal masses and hypodense areas in the white matter. But in 20% of cases, granulomas can coalesce, and previous articles have reported isolated thickening of the dura. This appears as an extracerebral iso- or hyperdense mass markedly enhanced on postcontrast CT and most often located over the convexity, although an infratentorial location is possible [3, 6–14]. This can mimic meningioma-en-plaque, but an appearance quite similar to the one in our study has also been observed with extensive involvement of the tentorium and posterior falx [15]. Angiograms can show an avascular mass [7, 9, 10, 13].

To our knowledge, only one MR report [8] of granulomatous dural/subdural involvement has been published, depicting variable appearances on T2-weighted sequences: hyper-, iso-, but also hypointense signal areas relative to cerebral cortex. Peripheral edema may be seen. A hypointensity with hyperintense edges, as observed in our case, has never been described; nevertheless, further studies are necessary to determine the value of MR in differentiating the appearances we describe in our cases from neurosarcoïdosis.

Moreover, no extracerebral lesion was observed in our patients. Serum angiotensin-converting enzyme levels and gallium brain scans were normal. The diagnosis of neurosarcoïdosis is definitely excluded by the absence of characteristic noncaseating granuloma on biopsies. Similar CT observations with diffuse dural infiltration occur in hematogenous disorders, especially lymphomas and myelofibrosis. To our knowledge, no MR report on dural lymphoma has appeared, but clinical presentation in those cases is quite different and microscopic observations disclose the cellular infiltration [16, 17].

Dural carcinomatosis can also appear on CT as crescentic thickened and enhanced dura mater. It is a rare manifestation of metastatic disease, but 15 isolated cases have been reported. A recently reported MR appearance [18] consisted of intermediate signal on both T1- and T2-weighted sequences, differing from the findings in our study. Thickening of dura may also be extensive, involving the convexity and skull base, but microscopic examination shows infiltration of dura by a fibrous vascular stroma containing intravascular tumor nests.

Late syphilis, a well-known cause of hypertrophic pachy­meningitis, is described in the earlier literature [3, 19]. The clinical presentation is similar to that in our cases, with chronic severe headaches, vertigo, ataxia, and various cranial nerve palsies correlating with compression of neighboring structures by the thickened dura mater. In the isolated case of Moore et al. [19], CT demonstrated dural enhancement with hypertrophy of the tentorium and falx indistinguishable from that in our cases. However, in our patients, all syphilis tests were negative and classical syphilitic endarteritis was never seen in our microscopic observations.

Rarely, fibromas can involve dura mater: the MR-pathologic correlation in case 1 suggests that the central area of decreased signal intensity on the T2-weighted sequence is due to the dense fibrous tissue, whereas the hyperintense signal of the edges corresponds to an inflammatory infiltrate of lymphocytes and plasma cells. Thus, given the MR appearance, fibromatoses diseases of the dura must be considered.

There are few reports of intracranial fibromas. They appear most often as well-circumscribed intracerebral masses related or not related to the meninges [20–22]. Extensive leptomeningeal fibrosis can also be present [23]. Intracranial fibromas can engulf and thicken the tentorium with marked CT enhancement [20]. Excepting pathologically confirmed infiltration of the underlying brain, CT features are very similar to those in our cases, though unilateral. Masses are often avascular on angiograms [20–22].

To our knowledge, MR of extracranial fibromatosis has not been reported except in neurofibromatosis, where neurofibromas present varied signals on T2-weighted sequences with, in some cases, central areas that can also display decreased signal intensity corresponding to fibrous components [24–26]. Aggressive fibromatosis has been shown by Aisen et al. [27] to have such decreased signal intensity on both T1- and T2-weighted images. But never has such peripheral high signal been described. This MR aspect, observed in case 1, therefore may be very useful in the differential diagnosis from other fibrous lesions. Microscopically, these lesions are moderately cellular, with spindle cells among a collagenous, slightly vascular stroma. Contrary to our observations, no similar infiltrates of lymphocytes and plasma cells are found, even though scattered lymphocytes can be seen [20–22, 28].

From a pathologic standpoint, the association of meningothelial cells with plasma cell–lymphocytic infiltrates in a collagenous stroma has been individualized as “plasma-cell granuloma” or “meningioma with conspicuous plasma cell components” [29–32]. These features could be compared with those in our cases, except only some meningothelial cells are seen
and, as with classical meningioma, distant dura mater involvement is lacking in these cases; also, radiologic findings are similar to those of meningiomas.

In two of our cases, the dural sinuses were narrowed or obliterated. This pattern probably is only related to nonspecific involvement of dura mater; indeed, this aspect has already been described with other infiltrative dura mater processes—in myelofibrosis [18], abnormal cells can invade sinuses. In neurosarcoïdosis, involvement of leptomeninges can be complicated by sinus thrombosis; the superior sagittal, straight, or lateral sinuses or the vein of Galen can then appear variously narrowed or occluded [13, 33]. Narrowing or occlusion of venous sinuses is seen in syphilitic or idiopathic pachymeningitis in relation to thickened sinus walls or obliteration by granulomatous pachymeningitis [3–5, 19].

Dural thickening on CT has been described by some authors [1, 34], with a hypointense signal area in one MR study [34] somewhat similar to the appearance in our cases, which the authors attributed to chronic venous sinus thrombosis. This raises the possibility that venous sinus thrombosis could lead to localized dural thickening.

In case 2, extensive hypodensities of the white matter were noted and were associated with mental deterioration. Diffuse periventricular hypodense areas in the white matter have been reported with patients suffering from neurosarcoïdosis; mental alterations and dementia can be present also. It is possible that our findings could result from involvement of the small blood vessels by granulomatous tissue (with stenosis or obliteration) [35, 36]. The absence of arteritis in our cases renders this hypothesis unlikely. Thus, the white-matter hypodensities may be related to the narrowing of dural sinuses and represent ischemic changes [37, 38]. In the same patient, the hydrocephalus (probably linked to obstruction of CSF drainage) could be a complication in the course of the disease.

Thus, the idiopathic hypertrophic pachymeningitis reported here appears to be unrelated to any known disease process. Some preceding reports are likely to correspond to similar observations. Our patients showed a good response to steroid therapy, particularly their headaches, but steroid therapy did not seem to arrest the clinical progression and the patients became steroid-dependent. A decrease in dural thickening on CT in case 3, correlating with a small-sized lesion on MR, was perhaps related to therapy; nonetheless, the clinical progression did not seem to be arrested. Bearing in mind parallel cases of neurosarcoïdosis unchecked by steroid therapy but improved by radiotherapy [39, 40], our two patients so treated did not show any durable improvement. Azathio­prine therapy might lead to clinical improvement; however, at this time it is still too early to tell.

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