

## Case Report

# Unusual Imaging Appearance of an Intracranial Dermoid Cyst

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**Summary:** Intracranial dermoid cysts have characteristic CT and MR imaging findings that generally make preoperative diagnosis straightforward. Enhancement of uncomplicated intradural dermoid cysts on CT or MR studies has been reported but is rare. We present a case of a posterior fossa dermoid cyst that was not only hyperattenuating on CT scans but also contained a mural nodule with clear evidence of enhancement on MR images.

Typical dermoid cysts are well-circumscribed fat-density masses with no associated contrast enhancement (1); rarely, they may appear hyperattenuating on CT scans (2–7). Dermoid cysts arise from the inclusion of embryonic ectoderm into the neural tube during the 5th to 6th weeks of fetal life and typically occur in the midline (1, 8). Intracranial dermoids are rare lesions, with the posterior fossa being the least common site of occurrence (0.04% to 0.7% of primary intracranial tumors) (1). These lesions usually present in the third decade of life with a long history of vague symptoms predominated by headache. Some are associated with seizures, focal neurologic deficits, and episodes of aseptic meningitis. Enlargement has been shown to be due to a combination of glandular secretions and epithelial desquamation. Rupture of these cysts either spontaneously or at surgery results in a chemical meningitis that may be severe, leading to vasospasm, infarction, and even death (8).

### Case Report

An 18-year-old girl had an 8-week history of occipital headaches, which were not particularly suggestive of raised intracranial pressure. During the preceding few weeks she had also experienced deteriorating vision in the right eye, loss of balance, frequent falls, loss of weight, anorexia, and depression. On examination, she had markedly decreased visual acuity in the right eye of 6/60 with marked papilledema in both optic disks, reduced corneal sensation, a partial right sixth cranial nerve palsy, plus left gaze and upbeat nystagmus. The lower

cranial nerves were normal. Peripheral examination revealed increased tone and brisk reflexes in all her limbs and an ataxic gait.

Pre- and postcontrast CT scans showed a  $7.5 \times 6.0 \times 5.5$ -cm hyperattenuating midline posterior fossa lesion in the region of the cerebellar vermis associated with obliteration of the fourth ventricle and acute obstructive hydrocephalus (Fig 1A). The lesion was well circumscribed, predominantly cystic, and contained a small posterior heterogeneous mural nodule with calcification. No definite nodular or mural contrast enhancement could be seen on the CT studies. MR imaging showed the lesion to be markedly hyperintense on T1-weighted images and extremely hypointense on T2-weighted studies (Fig 1B). Noncontrast images showed a posterolateral mural nodule with clear evidence of central enhancement in the region of serpiginous flow voids (Fig 1C and D). Sagittal images revealed marked herniation of the cerebellar tonsils (Fig 1E). The mass was avascular at digital subtraction angiography.

A midline low occipital craniotomy was performed. On opening the dura and midline arachnoid, the lesion was immediately entered, and thick, light-brown fluid with the consistency of caramel sauce was expressed. Deeper exploration of the cavity revealed hairs and thick sebaceous material, consistent with a dermoid cyst. Although the cyst wall was friable and fragmented at surgery, the lesion was completely excised without complication. The patient made a satisfactory postoperative recovery and was discharged 9 days after admission. At 2 months, her headaches and ataxia had completely resolved, although vision in her right eye was still at 6/60 with evidence of optic atrophy. A follow-up MR imaging examination at 8 months showed no recurrence of the lesion.

### Discussion

On the basis of the CT and MR imaging appearances (mural nodular enhancement and cystic attenuation/signal intensity suggestive of blood products), hemorrhage into a cerebellar hemangioblastoma, cavernous hemangioma, or an atypical dermoid cyst were considered the most likely differential diagnoses, with the understanding that the imaging appearances were unusual for all these lesions. Angiography effectively excluded a cerebellar hemangioblastoma, as these lesions virtually always have an intensely hypervascular mural nodule with prolonged vascular staining (1).

The signal intensity of the cystic contents on MR images remained consistent with either subacute hemorrhage or the fatty or partially liquified cholesterol-rich material seen in dermoids; however, the presence of an enhancing mural nodule and absence of a midline posterior occipital dermal sinus tract (characteristically seen with posterior fossa dermoids) made a dermoid cyst seem less likely (8). On CT scans, dermoids are usually rounded,

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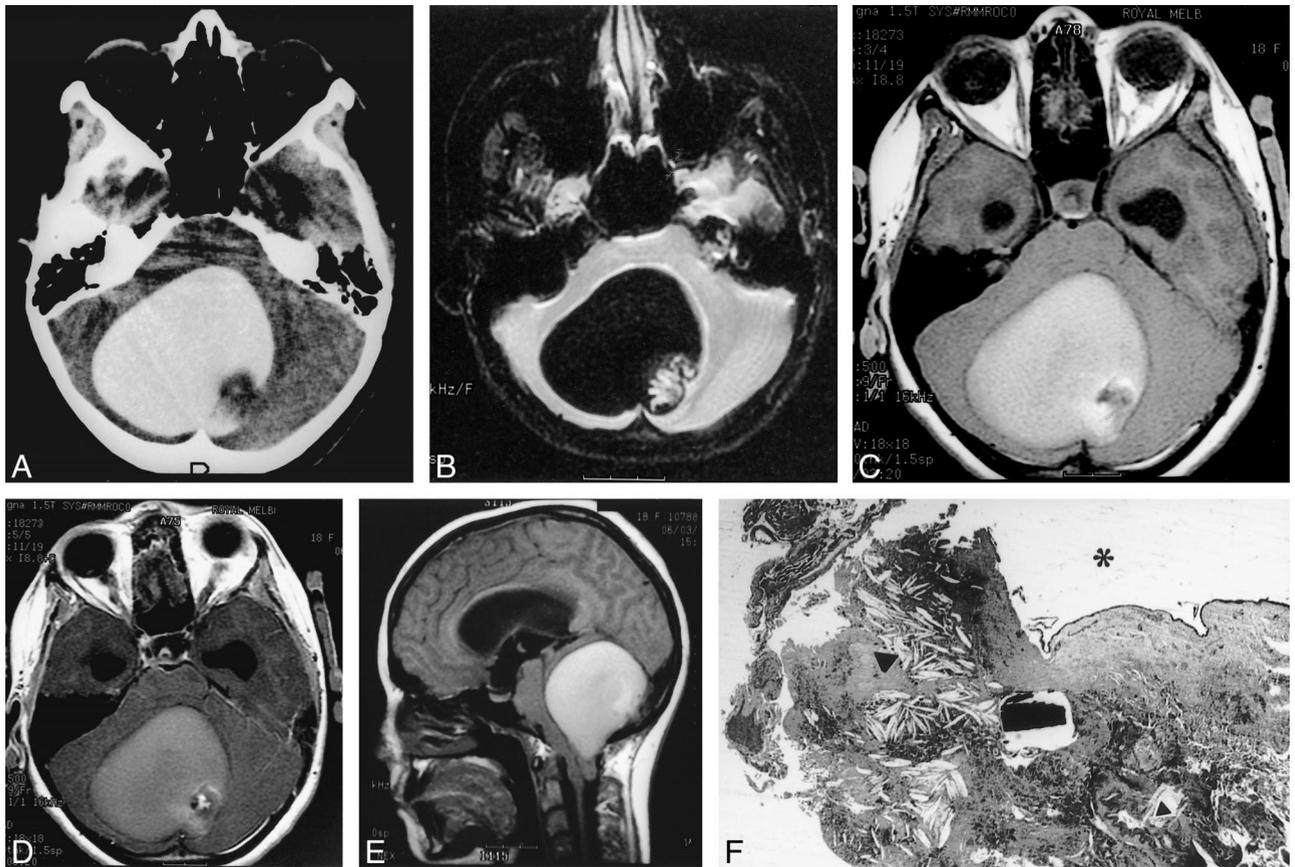


FIG 1. 18-year-old girl with occipital headaches of 8 weeks' duration.

A, Unenhanced CT scan shows a hyperattenuating posterior fossa lesion with posterior mural nodule.

B, Gradient-echo MR image shows marked hypointensity with a frondlike mural nodule.

C and D, Pre- (C) and postcontrast (D) T1-weighted MR images show hyperintensity of the cystic contents and clear evidence of contrast enhancement of the mural nodule.

E, Sagittal unenhanced T1-weighted MR image shows marked cerebellar tonsillar herniation through the foramen magnum and obliteration of the fourth ventricle.

F, Section of cyst wall (lumen indicated by asterisk) shows a lozenge-shaped deposit of calcium with a clear space centrally, numerous cholesterol deposits (large black arrowhead), and a leash of vessels (small black arrowhead) (hematoxylin-eosin, original magnification  $\times 25$ ).

well-circumscribed, extremely hypodense lesions with a Hounsfield unit of  $-20$  to  $-140$ , in keeping with their lipid content. They are never associated with vasogenic edema and only rarely cause hydrocephalus. Peripheral capsular calcification is frequent. Enhancement after contrast administration is rare but has been reported (1, 8, 9).

On MR images, dermoids are typically hyperintense on T1-weighted images but vary from hypo- to hyperintense on T2-weighted studies. Again, there is usually no associated vasogenic edema or contrast enhancement. Serpiginous hypointense elements may be seen if the lesion contains hair. Mural calcification can sometimes be identified. On both CT and MR images, fat-density droplets may be seen throughout the subarachnoid space and in the ventricular system if rupture of the cyst has occurred (1, 8, 9).

Dermoids presenting as hyperattenuating lesions on CT studies are extremely rare and, as such, present a diagnostic challenge. To our knowledge, only seven cases have been reported in the English-lan-

guage literature, and none had an enhancing mural nodule. In each of these cases, other lesions, such as hemorrhagic tumor, hematoma, or meningioma, were considered more likely on the basis of the initial CT imaging findings (2–7). On MR imaging, at least one of these lesions was markedly hypointense on T1- as well as T2-weighted images, suggesting that microcalcification was responsible for the CT hyperattenuation (7). Our lesion was markedly hyperintense on T1-weighted images, which is common on MR studies, but hyperattenuation is virtually never seen on CT scans.

Interestingly, all of the reported CT hyperattenuating dermoids have occurred in the posterior fossa, with no lesions ever having been identified supratentorially. We are unaware of a pathologic explanation for this.

Pathologically, the hyperattenuation seen on CT scans is thought to be due to a combination of saponification of lipid or keratinized debris with secondary microcalcification in suspension (also seen as capsular deposits in some lesions), partially liq-

unified cholesterol, high protein content, and hemosiderin or iron calcium complexes relating to previous episodes of intracystic hemorrhage. Note that fat per se is not found in dermoids as this is of mesodermal origin. Breakdown products of hair and secretions of sweat and sebaceous glands result in an oily fluid containing lipid metabolites (8, 10). Sections of the cyst wall in our patient showed a squamous epithelial lining with an occasional distorted pilosebaceous unit within fibrous tissue. Focal deposition of calcium was also noted as well as aggregates of cholesterol clefts (Fig 1F). A leash of somewhat distorted vessels was also present in one section. The cystic contents contained keratin, in which hair shaft remnants could be identified.

In summary, we have reported a case of a CT hyperattenuating posterior fossa lesion that showed evidence of enhancement within a mural nodule on MR images and was proved pathologically to be a dermoid cyst. Although this is a hitherto unreported combination of imaging features, separately they are both recognized as rarely being associated with these lesions and should not prevent the diagnosis

of dermoid once important differentials have been excluded.

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