
Occipital Dermal Sinus Associated with Dermoid Cyst in the Fourth Ventricle

Sotaro Higashi, Kenji Takinami, and Junkoh Yamashita

Summary: MR was helpful in demonstrating the spatial relationship between a dermoid cyst in the fourth ventricle and an occipital dermal sinus. T1-weighted images clearly showed the mixed hypointense mass in the fourth ventricle and above the vermis, which appeared to be connected to an occipital dermal dimple.

Index terms: Brain, abnormalities and anomalies; Brain, ventricles; Dermoid cyst

A congenital dermal sinus is a tract lined by epidermis forming a potential communication between the skin and the deeper tissues within the central nervous system. Occipital dermal sinuses associated with dermoid cysts in the fourth ventricle are rare.

Case Report

A 25-year-old woman had had a dimple with thickening of the scalp in the midoccipital region since birth. Tremor of her right hand had been present for 14 years and was gradually worsening. There was no clinical history of meningitis. Neurologic examination on admission showed horizontal nystagmus, tremor of the right hand, and right cerebellar ataxia. Plain skull radiographs showed an oval bony defect at the lambda and another small bony defect in the midoccipital region (Fig 1). Precontrast computed tomography showed a large, low-density lesion in the midline of the posterior fossa without hydrocephalus. Postcontrast computed tomography showed no enhancement. Sagittal and oblique coronal T1-weighted magnetic resonance (MR) images clearly showed the mixed hypointense mass lesion in the fourth ventricle and above the vermis, which appeared to be connected to the occipital dermal dimple. The straight sinus along the dermoid cyst above the vermis was clearly demonstrated (Fig 2A–C). The vermis was markedly atrophic and the dorsal brain stem was compressed (Fig 2 and 3). A suboccipital craniotomy was performed with the patient sitting. The tumor protruding into the cisterna magna from the fourth ventricle had a shiny pearly surface, suggesting an epidermoid cyst. The

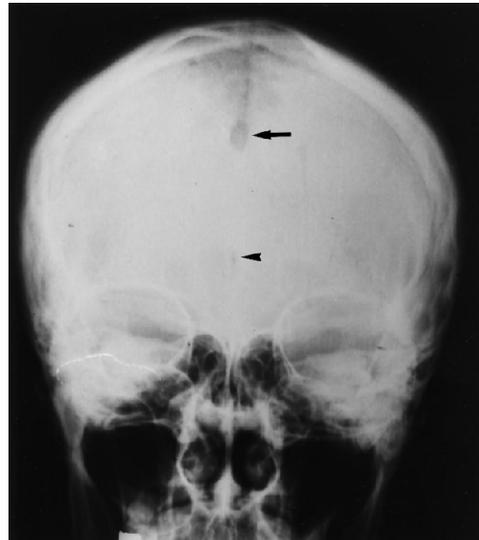


Fig 1. Plain skull radiograph shows an oval bony defect at the lambda (arrow) and another small bony defect (arrowhead) in the midoccipital region.

inside of the cyst was filled with flaky yellow material and hair. The tumor in the fourth ventricle was totally removed except for a thin capsule adhering to the floor of the fourth ventricle. The brain stem seemed compressed but not invaded by the tumor, as surgery through the translucent capsule of the tumor showed the floor of the fourth ventricle to have a smooth surface. The cavity was washed thoroughly with saline. A small orifice extending from the fourth ventricle into the quadrigeminal cistern was observed just posterolateral to the aqueduct, which was also clearly shown by the sagittal MR image (Fig 2B). The superficial part of the dermal sinus including the cutaneous lesion was removed, but the sinus tract extending under the skull and the dermoid cyst above the vermis were left intact (Fig 2D).

Histologic examination of the cyst wall revealed that the tumor was a dermoid cyst consisting of stratified squamous epithelium, hair follicles, and keratin debris. Sebaceous and sweat glands were not found, which indicated

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From the Department of Neurosurgery, School of Medicine, Kanazawa University, Japan.

Address reprint requests to Sotaro Higashi, MD, Department of Neurosurgery, School of Medicine, Kanazawa University, 13-1 Takara-machi, Kanazawa 920, Japan.

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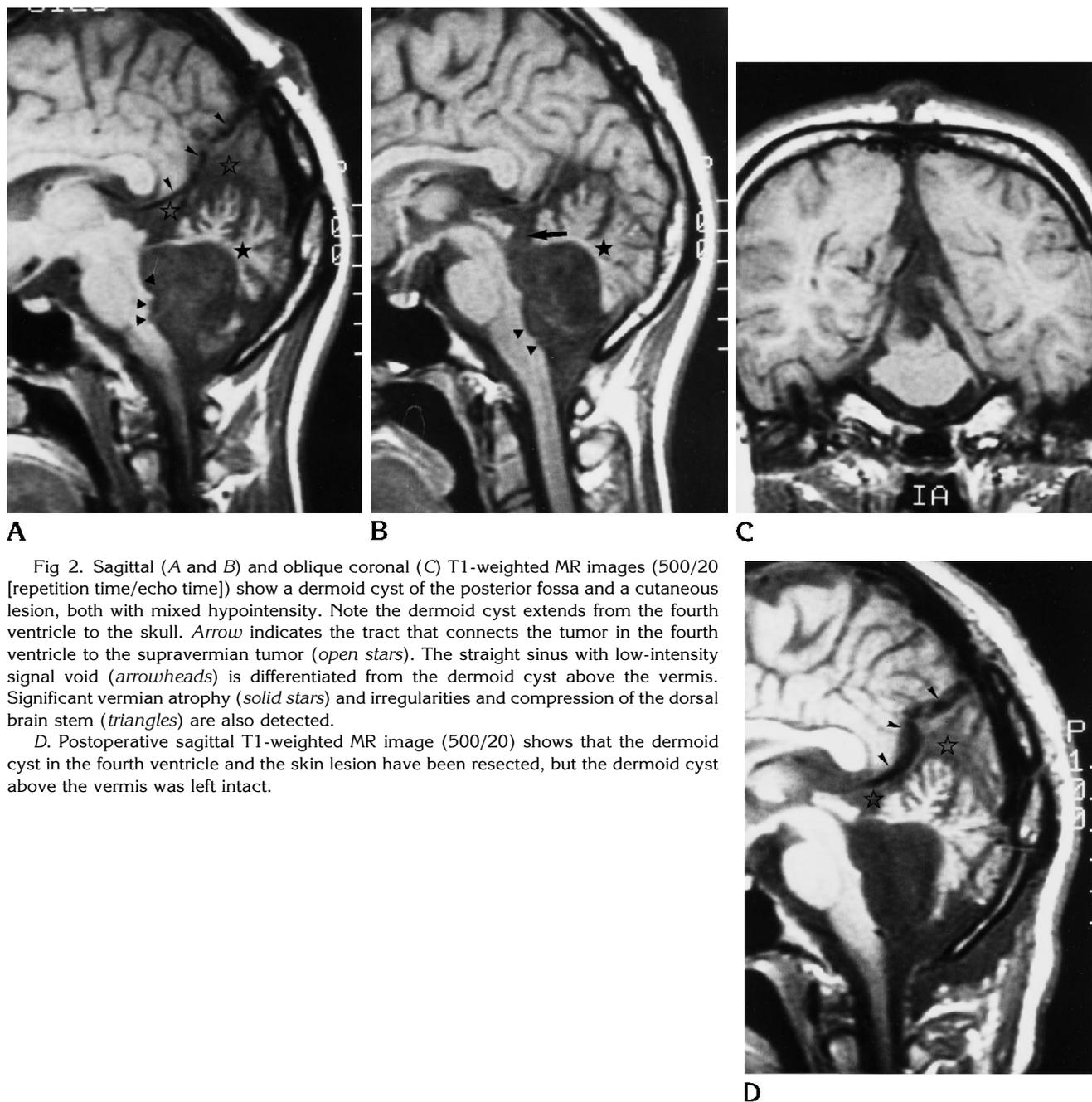


Fig 2. Sagittal (A and B) and oblique coronal (C) T1-weighted MR images (500/20 [repetition time/echo time]) show a dermoid cyst of the posterior fossa and a cutaneous lesion, both with mixed hypointensity. Note the dermoid cyst extends from the fourth ventricle to the skull. Arrow indicates the tract that connects the tumor in the fourth ventricle to the supravermian tumor (*open stars*). The straight sinus with low-intensity signal void (*arrowheads*) is differentiated from the dermoid cyst above the vermis. Significant vermian atrophy (*solid stars*) and irregularities and compression of the dorsal brain stem (*triangles*) are also detected.

D. Postoperative sagittal T1-weighted MR image (500/20) shows that the dermoid cyst in the fourth ventricle and the skin lesion have been resected, but the dermoid cyst above the vermis was left intact.

that the dermoid cyst may have contained less lipid or liquefied cholesterol (Fig 4). The patient had a good postoperative recovery without chemical or bacterial meningitis.

Discussion

We describe a rare case of an occipital dermal sinus associated with a dermoid cyst in the fourth ventricle and above the vermis. Congenital dermal sinuses can occur anywhere along the craniospinal axis. Wright (1) collected 127

cases of congenital dermal sinuses from the literature: 72 lumbosacral, 30 occipital, 12 thoracic, 2 cervical, 1 sacrococcygeal, and 10 of various sites. French (2) reviewed the cases of intracranial congenital dermal sinuses and found that 85% of the dermal sinuses were located near the external protuberance of the occipital bone, 11% at the nasion, and 5% in the posterior parietal area. Eighty-nine percent of the dermal sinuses were associated with an in-

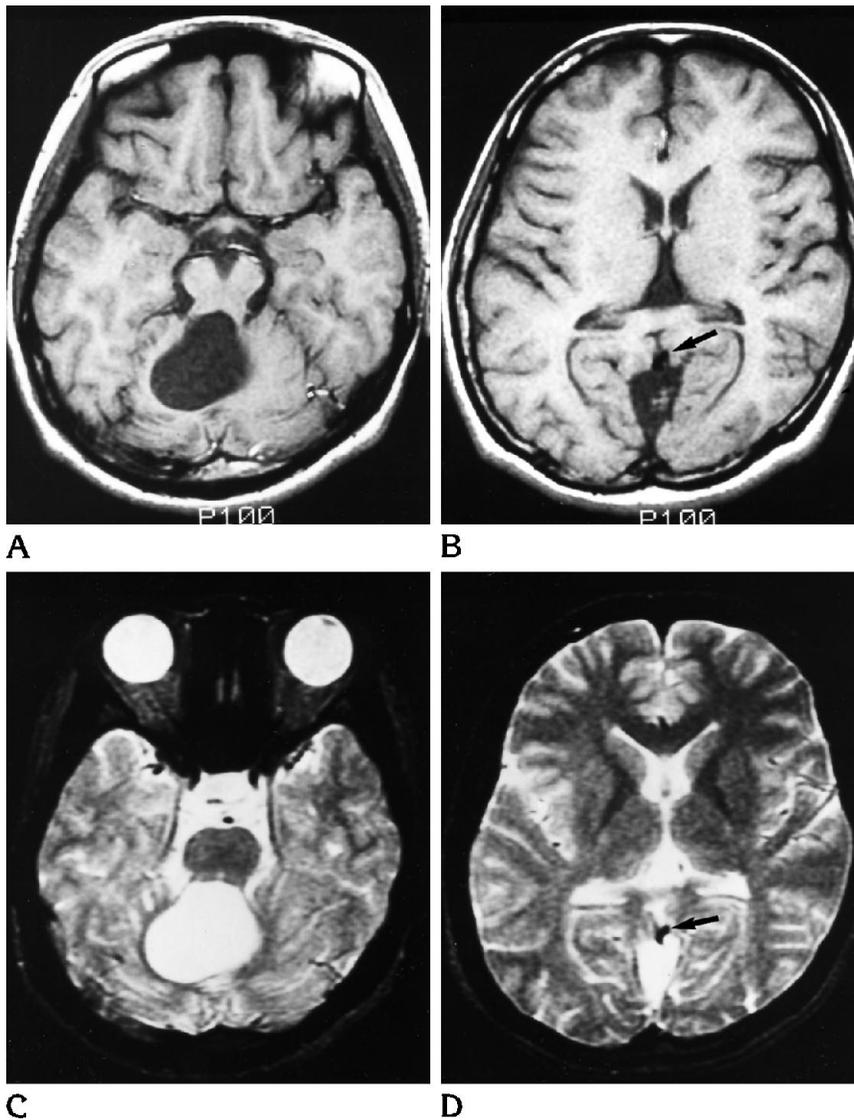


Fig 3. Axial T1-weighted MR images (500/20) (A and B) and corresponding T2-weighted MR images (2500/80) (C and D). The MR appearance of the dermoid cyst is of mixed hypointensity on T1-weighted sequences and hyperintensity on T2-weighted sequences. *Arrows* indicate the straight sinus.

clusion tumor. French also reported that all these tumors were dermoid cysts except for three that were classified as epidermoid cysts, and that the tumor was extradural in 18% of cases and subdural or deeper in 82%. Although approximately one third of intracranial dermoid cysts without a dermal sinus are in the fourth-ventricular region (3), we found only two cases of a dermal sinus associated with a dermoid cyst in the fourth ventricle reported (4).

Epidermoid cysts are usually hypointense on T1-weighted sequences because of the presence keratin and cerebrospinal fluid; however, those with high lipid content can be hyperintense on T1-weighted sequences. Epidermoid cysts are relatively hyperintense on T2-weighted sequences, but those that are lipid

containing can be hypointense relative to cerebrospinal fluid (5). MR findings of dermoid cysts are more variable. Smith et al (6) analyzed the MR findings in seven patients who had pathologically proved ruptured dermoid cysts and reported that the signal intensity of the solid mass was low on T1-weighted sequences and inhomogeneously high on T2-weighted sequences in all cases. The relative proportions of the elements of a dermoid cyst can vary. In general, it has thick, viscous, greenish brown fluid composed of lipid metabolites and liquid cholesterol from decomposed epithelial cells. These liquid products explain hyperintensity on T1-weighted sequences, with lower signal on T2-weighted images (5, 6). In our case, the MR appearance agreed with that reported by Smith et al: mixed



Fig 4. Photomicrograph of the surgical specimen shows the lining of the cyst consists of stratified squamous epithelium (*small arrows*) with hair follicle (*large arrow*) and keratin debris (*star*). Sebaceous and sweat glands are not found, indicating that the dermoid cyst may contain less lipid or liquefied cholesterol.

hypointensity on T1-weighted sequences and hyperintensity on T2-weighted sequences (Fig 3), simulating epidermoid cysts. Generally speaking, differentiation between dermoid cysts and epidermoid cysts is not easy with radiologic imaging and is confirmed only by histologic examination. This is true especially when most of the contents of the dermoid cyst are keratin and cerebrospinal fluid, with less lipid or liquefied cholesterol.

Atretic cephaloceles are lesions that need to be distinguished from dermal sinuses with scalp dermoid cysts. Scalp dermoid cysts do not enhance on postcontrast computed tomography, whereas atretic cephaloceles do enhance (7).

Preoperative anatomic delineation and evaluation of cystic contents with MR helps to determine the surgical approach. In our case, direct sagittal and oblique coronal MR clearly visualized the entire dermoid cyst, the mass effect on the brain stem, and the vermian atrophy. These effects can be caused by longstanding, slowly progressive compression by the dermoid cyst. In our case, the cyst contents were suspected of containing less lipid or liquefied cholesterol, indicating a diminished possibility of postoperative aseptic meningitis.

Occipital dermal sinuses are benign lesions that are usually discovered because of recurrent meningitis, cerebellar abscess, or infection in the cutaneous opening (8). Postoperative aseptic meningitis, caused by spillage of cyst contents, is also a common problem. Ideal treatment of this benign lesion is complete surgical removal followed by thorough irrigation. In our case, the deep-seated sinus tract and the dermoid cyst above the vermis were not removed, as MR had shown that the dermoid cyst above the vermis probably was attached firmly to the straight sinus and the thin cyst capsule was closely adherent to the floor of the fourth ventricle. In such cases, subtotal removal is justified if there is no evidence of infection in the cutaneous opening nor in the associated tumor.

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