

CEREBELLAR DERMOID CYST WITH CONTRAST ENHANCEMENT MURAL NODULE: CASE REPORT

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SUMMARY – Typical dermoid cysts are well-circumscribed fat-density masses with no associated contrast enhancement; rarely, they may appear hyperattenuating on CT scan. CT hyperattenuating dermoid cyst (CHADC) is very uncommon, with only nine case reports in the literature update, which occurs exclusively in the posterior fossa. CHADC with mural nodule is extremely rare and, to the best of our knowledge, only two cases have been documented previously in the literature. A 49-year-old farmer had a 2-month history of occipital headaches, which were not suggestive of raised intracranial pressure. During the last month, he experienced loss of balance, frequent falls, anorexia and loss of weight. Magnetic resonance imaging (MRI) showed a huge mass from the tentorium to the foramen occipitale magnum with obliteration of the fourth ventricle; the lesion was well circumscribed. We completely removed the tumor and postoperative MRI showed no residual tumor. Epidermoid tumors with enhancing mural nodule on MRI and with hyperattenuating lesion on CT are extremely rare. Dermoid cysts are never associated with edema and extremely rarely cause obstructive hydrocephalus. MRI investigations are mandatory to diagnose these cases. The best curative treatment is total removal of the lesion.

Key words: *Cerebellar neoplasms – radiography; Cerebellar neoplasms – surgery; Cranial fossa, posterior – radiography; Cranial fossa, posterior – surgery; Dermoid cyst – radiography; Dermoid cyst – surgery; Case reports*

Introduction

Typical dermoid cysts are well-circumscribed fat-density masses with no associated contrast enhancement¹; rarely, they may appear hyperattenuating on computed tomography (CT) scans¹⁻⁷. Dermoid cysts originate from the totipotential ectodermal cells, which remain within the developing neural tube between the third and fifth week of gestation. 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. More than 50% of these tumors are diagnosed in childhood or early adolescence. Dermoid cyst, a congenital lesion, is classified into a group named ‘inclusion tumors’, besides the more prevalent tumors like epidermoids and hamartomas. Dermoid cyst is the least common one in this group⁸. Dermoid cysts present in most cases between third and fourth decade of life, with a long history of vague symptoms predominated by severe headache. Chemical meningitis due to rupture of dermoid cyst either spontaneously or at surgery may cause vasospasm, infarction, and even death. Logue and Till⁹ have classified posterior fossa dermoid cysts into 4 groups, depending on whether they are extradural or intradural, and on the degree of development of the dermal sinus, whether absent, partial, or complete: (1)

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extradural dermoid cyst with a complete dermal sinus; (2) intradural dermoid cyst (with no dermal sinus); (3) intradural dermoid cyst with an incomplete dermal sinus; and (4) intradural dermoid cyst with a complete dermal sinus. Our case was of type 2, according to this classification. CT hyperattenuating dermoid cyst (CHADC) is very uncommon with only nine case reports in the literature update, which occurs exclusively in the posterior fossa. CHADC with mural nodule is extremely rare and, to the best of our knowledge, only two cases have been documented previously in the literature, so our case is the third one.

Case Report

A 49-year-old farmer presented with a 2-month history of occipital headaches, which were not suggestive of raised intracranial pressure. During the last month he experienced loss of balance, frequent falls, anorexia, loss of weight, and also showed some signs of depression. On neurological examination, he had ataxic gait and increased tone in all his limbs. Horizontal nystagmus on gaze to the right and very poor coordination on the finger to nose test were observed, while the heel-shin test was not as poor as the finger

to nose test. Ophthalmologic examination revealed reduced visual acuity to 3/10 on the left eye and 4/10 on the right eye, with unilateral stage II papillary edema at funduscopy. Preoperative laboratory tests were within the normal range. In sitting position, incision was made to C2 and suboccipital osteoplastic craniotomy was performed. The dura was opened in the Y fashion. When the first burr hole was made, a dose of 125 mL of mannitol was administered intravenously. A tumor which displaced the cerebellum anterolaterally was found. The posterolateral surface of the tumor was adherent to the overlying dura matter, but there was no aggression to the brain tissue. MRI showed a huge mass from the tentorium to the foramen occipitale magnum with obliteration of the fourth ventricle; the lesion was well circumscribed (Figs. 1 and 2).

The tumor contained a small posterior heterogeneous mural nodule without calcification. Operation under surgical microscope was performed (magnification X3.2-4.1). When the dura was opened, the lesion was immediately entered and fluid of caramel sauce consistency was expressed. A thin capsule had

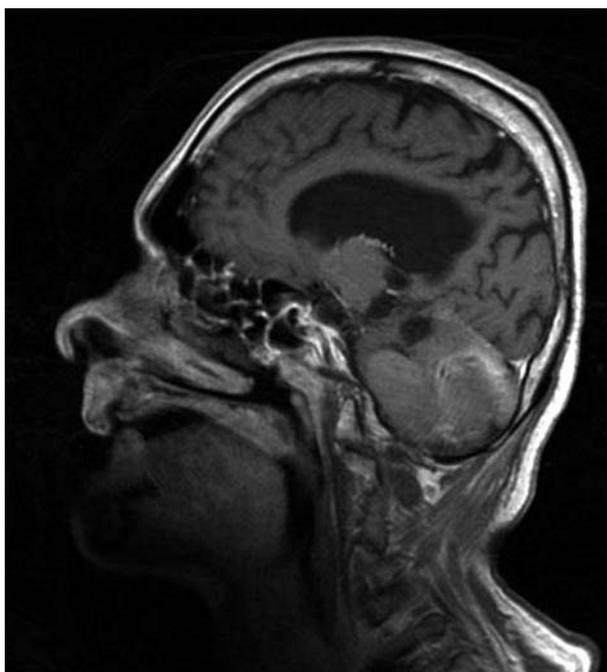


Fig. 1. Magnetic resonance imaging, sagittal section.

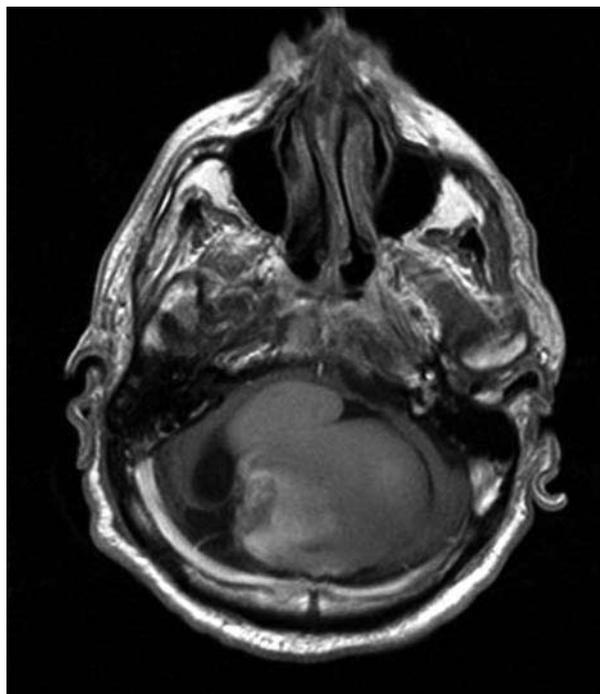


Fig. 2. Magnetic resonance imaging, axial section: hyperintensity of the cyst contents and clear evidence of contrast enhancement of the mural nodule.

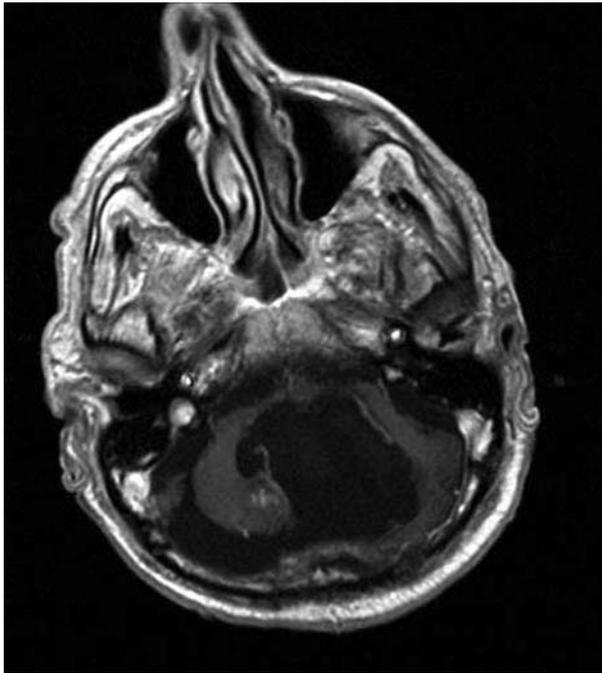


Fig. 3. Postoperative magnetic resonance image, axial section: no residual tumor was left.

surrounded the mass, which was removed gently as much as possible. The lesion was completely excised without any complication. During the surgery, the evoked potentials were normal. The dura was closed water tight and there was no need for duraplasty. Besides that, the adhesions were very close between the floor of the fourth ventricle and the tumor; resection of the mass was total. The drain was removed on the second postoperative day. Post to operative laboratory tests were within the normal range. Also, postoperative function of cranial nerves was without any neurological deficit. Before patient discharge, control postoperative MRI with contrast was performed (Fig. 3). The patient made a satisfactory postoperative recovery and was discharged 10 days after admission to our department. His postoperative course was good. Histopathologic study confirmed the diagnosis of dermoid cyst (Fig. 4). At 3 months, his headaches, ataxia and nystagmus had completely resolved. A follow up MRI examination at 6 months showed no recurrence of the lesion.

Discussion

The age at presentation of this tumor is usually childhood and early adolescence. The origin of these

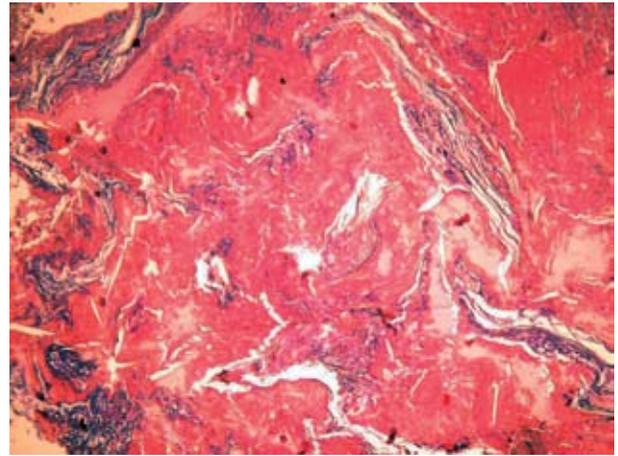


Fig. 4. Masses of amorphous keratin associated with the areas of dystrophy calcification in the lumen of the cyst (H&E, X100).

tumors is implantation of abnormal tissue during neural tube closure in the third to fifth week of embryonic life. We report on a posterior fossa dermoid tumor that was not only hyperdense on CT scan but also contained a mural nodule with clear MRI evidence of enhancement. On the basis of the CT and MRI appearances (mural nodular enhancement and cystic attenuation/signal intensity suggestive of blood products), hemorrhage into 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 cerebellar hemangioblastoma, as these lesions virtually always have an intensely hypervascular mural nodule with prolonged vascular staining¹. Epidermoid tumors with enhancing mural nodule on MRI and with hyperattenuating lesion on CT are extremely rare.

Dermoid cysts are never associated with edema and extremely rarely cause obstructive hydrocephalus. Our experience suggested that high protein, high calcium and low lipid contents were the main mechanism of increased CT attenuation for CHADC, along with slight male predilection. Malignant transformation is extremely rare and these lesions are slow growing. There are cases of posterior fossa dermoid tumors associated with Klippel-Feil syndrome reported in the literature. The best prognosis is associated with total microneurosurgical removal, whereas partial removal carries re-

markable freedom from recurrence. If a posterior fossa mass is seen on MRI or CT in patients with Klippel-Feil syndrome, dermoid cyst should be considered¹⁰.

In the revised World Health Organization classification, dermoid cysts are placed among the malformative or tumor-like lesions. Mortality and morbidity increase if chemical or bacterial meningitis develops, or if there are associated cerebellar abscesses¹¹. Otherwise, no recurrence after complete removal of the cyst has been reported in the literature¹². Dermoid cysts of the posterior fossa are uncommon. They usually manifest with cerebellar syndrome, intracranial hypertension, and sometimes with meningitis. The key examination remains MRI, while surgery remains the treatment of choice beyond any doubt.

References

- OSBORN AG. Diagnostic neuroradiology. St Louis: Mosby, 1944:635-6, 104-5, 533-57.
- BIZZOZERO L, TALAMONTI G, D'ANGELO VA, CASADEI GP, ARRIGONI GL, COLLICE M. Dermoid cyst mimicking hematoma in the posterior fossa. Clin Neurol Neurosurg 1992;94:61-3.
- CECCHINI A, PAZZOTTA S, PAOLETTI P, RONGNONE F. Dense dermoids in craniocervical region. J Comput Assist Tomogr 1983;7:479-83.
- DANAILA L, CARP N. Dermoid tumor of the fourth ventricle with hyperdense aspect demonstrated on CT: a case report. Neurol Psychiatry 1989;27:231-6.
- DROLSHAGEN LF, STANDEFER M. Dense dermoid cyst of the posterior fossa. AJNR Am J Neuroradiol 1991;12:317.
- GOH GJ, PAGE RD, NIXON TE. An unusual CT and MR appearance of a posterior fossa dermoid. Eur J Radiol 1995;20:46-7.
- LUNARDI P, RIZZO A, GUIDETTI G. Unusual CT-dense transtentorial epidermoid cyst. Neurochirurgia 1988;31:219-21.
- YOUSMANS JR. Youmans neurological surgery, Fifth Edition, Vol. 4. Philadelphia, PA: Saunders, 2004:4259-60, 1223-4, 3690-2.
- LOGUE V, TILL K. Posterior fossa dermoid cysts with special reference to intracranial infection. J Neurol Neurosurg Psychiatry 1952;15:1-12.
- KENNEDY PT, McAULEY DJ. Association of posterior fossa dermoid cyst and Klippel-Feil syndrome. AJNR Am J Neuroradiol 1998;19:195-6.
- DOUVOYIANNIS M, GOLDMAN DL, ABBOTT IR 3rd, LITMAN N. Posterior fossa dermoid cyst with sinus tract and meningitis in a toddler. Pediatr Neurol 2008;39:63-6.
- AKHADDAR A, JIDDANE M, CHAKIR N, EI HASANI R, MOUSTARCHID B, BELLAKHDAR F. Cerebellar abscess secondary to occipital dermoid cyst with dermal sinus: case report. Surg Neurol 2002;58:266-70.

Sažetak

CEREBELARNA DERMOIDNA CISTA S KONTRASTNIM POJAČANJEM MURALNOG ČVORIĆA: PRIKAZ SLUČAJA

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Tipične dermoidne ciste dobro su ograničene tvorevine intenziteta masti koje ne primaju kontrast i koje su rijetko kada hiperdenzne na kompjutorskoj tomografiji (CT). CT hiperdenzna dermoidna cista (CHADC) iznimno je rijetka te je dosad u literaturi opisano samo devet slučajeva, svi smješteni u stražnjoj jami. CHADC s muralnim nodulom je iznimno rijetka; prema našim saznanjima, dosad su u literaturi opisana samo dva slučaja. U ovom radu prikazujemo slučaj muškarca u dobi od 49 godina koji se žalio na dvomjesečne okcipitalne bolove bez znakova porasta intrakranijskog tlaka. Tijekom posljednjeg mjeseca često je padao zbog poremećaja ravnoteže, dok je zbog anoreksije smršavio. Magnetska rezonanca (MRI) je pokazala veliku, dobro ograničenu tvorevinu koja se protezala od tentorija do velikog zatiljnog otvora obliterirajući pritom četvrtu komoru. Tumor smo odstranili u cijelosti te MRI poslije operacije nije pokazao nikakvih rezidua bolesti. Na CT-u hipodenzni epidermoidni tumori s muralnim čvorićem koji prima kontrast na MRI su iznimno rijetki. Dermoidna cista nikad nije povezana niti s edemom i iznimno rijetko uzrokuje opstruktivni hidrocefalus. Slikovni MRI prikaz dijagnostički je u ovim slučajevima obvezatan za postavljanje dijagnoze. Najučinkovitija metoda liječenja je potpuno uklanjanje tvorevine.

Ključne riječi: *Mali mozak, tumori – radiografija; Mali mozak, tumori – kirurgija; Lubanjska jama, stražnja – radiografija; Lubanjska jama, stražnja – kirurgija; Dermoidna cista – radiografija; Dermoidna cista – kirurgija; Prikazi slučaja*