CASE REPORT

Atypical CT and MRI aspects of an epidermoid cyst

Aspects TDM et IRM atypiques d’un kyste épidermoïde

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Abstract We report a case of an unusual epidermoid cyst (EC) of the cerebellopontine angle that appeared hyperdense on computed tomography (CT) scanning, hyperintense on T1-weighted MR images and hypointense on T2-weighted magnetic resonance (MR) images. Diffusion-weighted imaging showed a hypointense lesion. We discuss imaging characteristics of ECs, explain the atypical findings in our case and confirm that the signal seen on diffusion-weighted images in the EC is related to a T2 effect.

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Introduction

Epidermoid cysts (ECs) are rare, benign, congenital tumors that represent 0.2-1% of all primary intracranial neoplasms [2,5,11,13]. They typically appear as well-defined lobulated masses hypodense on computed tomography (CT) and hypointense on T1-weighted images and hyperintense on T2-weighted images on magnetic resonance (MR) studies [8,9]. Many authors have described the diffusion-weighted features in typical ECs [1,2,4,9,10]. We report an EC of the cerebello-pontine angle (CPA) with an unusually dense appearance on CT scan and an unusual signal on MRI in a 39-year-old patient.

Case report

A 39-year-old woman presented with a 12-month history of headaches, dizziness, diploplia and walking difficulty with instability. Her medical history was positive for meningitis during childhood.

Neurological examination showed right static cerebellar syndrome, right vestibular syndrome and a deficit of the right ninth cranial nerve. CT scanning demonstrated a dense mass of the right CPA with no enhancement after intravenous contrast media (Fig. 1). On MR images, the
lesion was hyperintense on T1-weighted sequences (Fig. 2), and hypointense on T2-weighted sequences (Fig. 3). Contrast-enhanced T1-weighted images showed no appreciable enhancement. On the FLAIR and CISS 3D sequences, the signal was inhomogeneous (Figs. 4 and 5). Diffusion-weighted images depicted the lesion as hypointense (Fig. 6). The ADC was inhomogeneous, normal (0.710 × 10^{-3} mm^2/s equal to the ADC of the cerebellum parenchyma) with area of low diffusion (ADC = 0.587 × 10^{-3} mm^2/s) (Fig. 7). The imaging was thought to be most consistent with an unusual EC.

The cyst was approached using a lateral suboccipital craniectomy. After opening the dura, the tumor was visualized as a greenish multilobulated mass with a thin wall and contained debris. The mixed cranial nerves were surrounded by the tumor. The cyst was totally removed and the histopathological diagnosis was consistent with EC.

Figure 1 Noncontrast CT scan shows a dense extraaxial tumor in the right CPA.

Figure 2 T1-weighted image shows a hyperintense tumor of the right CPA.

Figure 3 T2-weighted image shows the tumor as hypointense.

Figure 4 FLAIR imaging shows the tumor as unevenly hypointense.

Figure 1 Coupe TDM sans produit de contraste mettant en évidence un processus expansif extra-axial hyperdense de l’angle ponto-cérébelleux droit.

Figure 2 Image axiale pondérée en T1 montrant une masse hyperintense à contours irréguliers de l’angle ponto-cérébelleux droit.

Figure 3 Image axiale pondérée en T2 montrant une masse hypointense de l’angle ponto-cérébelleux droit.

Figure 4 Image axiale pondérée en FLAIR montrant le signal hétérogène de la masse.
Discussion

ECs are benign dyssembryoplastic tumors caused during the third to fifth week of gestation by incomplete cleavage of the neural tissue from the cutaneous ectoderm [3,5,8,11]. They were first described by Lepreste in 1828 [11]. Acquired ECs caused by head trauma or lumbar puncture are rare and usually present as extradural masses [5,8].

ECs have a thin capsule of stratified, keratinized squamous epithelium. They are usually filled with white waxy material rich in cholesterol crystals mixed with cellular debris, which is the result of progressive desquamation and breakdown of keratin from the epithelial lining [2,3,5,11]. Their slow linear growth accounts for the late age of presentation (usually between the third and fifth decade of life) and the rather important size of the mass upon diagnosis [2,5,10,11].

They are most common in the CPA, the suprasellar region, the middle fossa and the quadrigeminal region [7–9].

The most frequent and suggestive symptoms at presentation in ECs of the CPA reflect seventh and eighth cranial nerve involvement. Involvement of the facial nerve is common and is most often represented by tic douloureux [8,9].

On CT scans, ECs appear heterogeneous, almost isoattenuationg to cerebrospinal fluid (CSF), and have characteristic irregular, lobulated margins [2,5,10-12]. Marginal calcifications and enhancement after contrast media administration are rarely seen.

In the typical cases, the diagnosis can be done on conventional T1- and T2-weighted images. However, some ECs present with a signal similar to that of CSF, making them hard to differentiate from an arachnoid cyst or with an unusual hyperintense signal compared to CSF on T1-weighted images. In these cases, sequences such as FLAIR, CISS 3D and diffusion-weighted imaging are very useful [2,5,6,8,10,11].

On FLAIR sequences, the signal is heterogeneous, hyperintense to CSF. On CISS sequences, ECs appear heterogeneous and hypointense compared to CSF. Because the CISS sequence provides high spatial resolution, multiplanar reconstructions and MR cisternography, it is an excellent tool to detect the exact tumor extension and to obtain precise delineation of the surrounding neurovascular structures [2,5,8,10,11]. ECs appear strongly hyperintense on diffusion-weighted images [1,2,4,12,14]. The origin of the hyperintense signal was attributed by Marin et al. [10] to the restricted diffusion of water molecules that are linked to the cholesterol molecules contained in these cysts, and in this case by a low ADC. This also suggests the solid nature of ECs. Shuda et al. measured the signal intensity and the ADC in ECs, normal cerebral tissue and CSF and found a 130% higher signal in ECs than that of normal cerebral tissue.
with a mean ADC higher than that of normal cerebral tissue ($1.197 \times 10^{-3}$ vs. $1.002 \times 10^{-3}$ mm$^2$/s). They suggested that the hyperintense signal of ECs on diffusion-weighted images is related to a T2 shine-through effect and not to restricted diffusion [3,7].

A few cases of hyperdense ECs have been reported [7,11-13]. On MRI, these lesions appear hyperintense on T1-weighted images and hypointense on T2-weighted images. Timmer et al. [13] explains these unusual features on CT and MRI by high protein concentration and high viscosity. He attributes the high signal intensity seen on T1-weighted imaging to the relatively high protein concentration of the cystic contents (> 90 mg/l). The low signal on T2-weighted images can be explained by the high viscosity of the fluid [13].

The hypointense signal with ADC values found in our case on diffusion-weighted images confirms that the signal of ECs on diffusion-weighted images is related to a T2 dark-through effect.

References