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Case Reports & Case Series

Co-existence of meningioma and epidermoid cyst in the posterior cranial Fossa

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ABSTRACT

We present a clinical case of combined tentorial meningioma and cerebellopontine angle epidermoid cyst (EC) in a 33-year-old woman. According to the literature, ours is the fifth case involving a combination of such tumours and the first in which the tumours were located in the posterior cranial fossa. Occurrence of a combination of brain tumours with different histological structures in the same patient is very rare. Nevertheless, the possibility of such a combination should always be considered, and thorough preoperative diagnostics are required to prevent complications and misdiagnosis. Currently, the combination of meningioma and EC is considered co-incidental. However, the proximity of the tumours described in the available reports does not exclude the possibility of one tumour's local influence on the other, which should be further studied.

1. Introduction

International studies have reported the occurrence of a combination of primary brain tumours of different histological natures in the same patient. The combined occurrence of glial tumours and meningiomas is the most frequently observed [1–3]. Additionally, various combinations of meningioma and epidermoid cyst (EC) with vestibular schwannoma, pituitary adenoma, gliomas of various anaplastic grades, and lipoma have been reported [1–6]. A combination of meningioma and EC is less commonly reported; only four cases have been reported in the English literature. Moreover, there are no previous reports on the occurrence of a combination of such tumours in the posterior cranial fossa (PCF).

We searched PubMed for reports published between 1970 and 2017 using the following keywords: meningioma, epidermoid cyst (along with terms, such as "and", "combined", "associated", and "coexistence").

Thomalske and Galow (1975) published the first report describing the surgical treatment of middle cranial fossa meningioma and clivus/cerebellopontine angle (CPA) epidermoid cyst (EC) [7]. Frazer and Victoratos (1979) described right orbit fibrosarcoma in a 3-year-old girl who underwent radiation treatment after tumour surgery. Thirty-three years after the initial diagnosis, the patient was diagnosed with suprasellar EC; 2 years later, she developed right frontal meningioma. The authors discussed the possible role of radiation therapy in the induction of intracranial mass lesions [8]. Kumar et al. (1992) described a tumour in the right frontal region in a 17-year-old woman whose histopathological findings revealed meningioma with EC [9]. Karekezi et al. (2016) published the surgical outcomes of a 37-year-old patient with atypical grade II right frontal lobe meningioma and right temporal lobe EC [10].

2. Case presentation

A 33-year-old woman presented to the clinic with complaints of severe headache, nausea, vomiting, severe dizziness, unsteady gait and progressive hearing loss in the left ear and was then admitted.

Her disease history showed initiation of progressive left ear hearing loss since approximately 1 year; she was followed up by an ENT specialist. In the previous 3 months, she also had headache, dizziness and unsteady gait. Therefore, she underwent brain magnetic resonance imaging (MRI), which revealed mass lesions in the left PCF, due to which she was referred to our clinic.

The patient's neurological status included left-sided hypoacusia (32 dB) and expressed coordination disorder.

MRI revealed a giant left-sided cerebellar tentorial meningioma with suprasubtentorial growth (the volume of the subtentorial part of the tumour significantly exceeded that of the supratentorial part) and relatively large left-sided CPA EC extending into the prepontine cistern,

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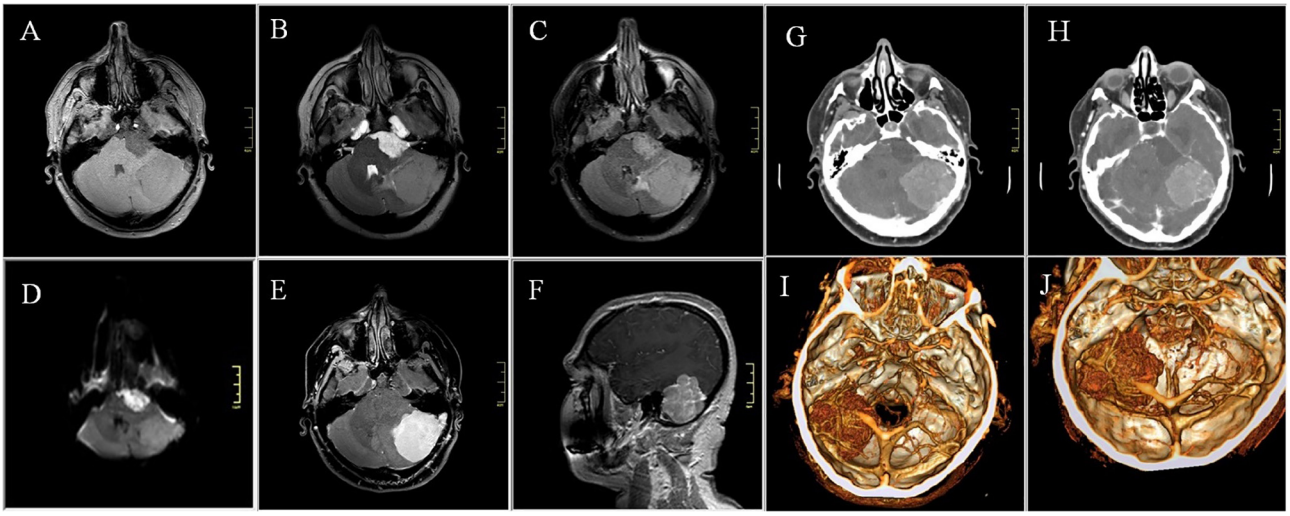

Fig. 1. Pre-operative magnetic resonance images (MRIs; **A–F**) and computed tomographic (CT) angiograms (**G–J**), showing a giant left-sided cerebellar tentorial meningioma with supra-subtentorial growth and a relatively large epidermoid cyst on the left side of the cerebellopontine angle, extending into the prepontine cistern, clivus and parasellar region. **A.** T1-weighted MRI. **B.** = T2-weighted MRI. **C.** Fluid-attenuated inversion recovery image. **D.** Diffusion-weighted imaging (DWI). **E.** T1-weighted MRI with gadolinium (Gd) enhancement, axial projection. **F.** T1-weighted MRI with Gd enhancement, sagittal projection. **G** and **H.** Native CT images. **I** and **J.** Three-dimensional CT reconstruction.

clivus and parasellar region (Fig. 1).

Pre-operative planning included computed tomography (CT) angiography, which revealed transverse sinus occlusion caused by giant tentorial meningioma (Fig. 1).

The tumours were removed using the left-sided extended retrosigmoid approach (trepanation above and below the transverse sinus). The subtentorial part of the meningioma was removed first, followed by the supratentorial part (Fig. 2). Furthermore, the tentorial area (meningioma matrix) and occluded sinus were resected. The meningioma removal grade was Simpson I. EC masses were then removed from CPA, prepontine cistern, clivus and parasellar region. Neurophysiological monitoring of facial nerve function using the Nim Response 3.0

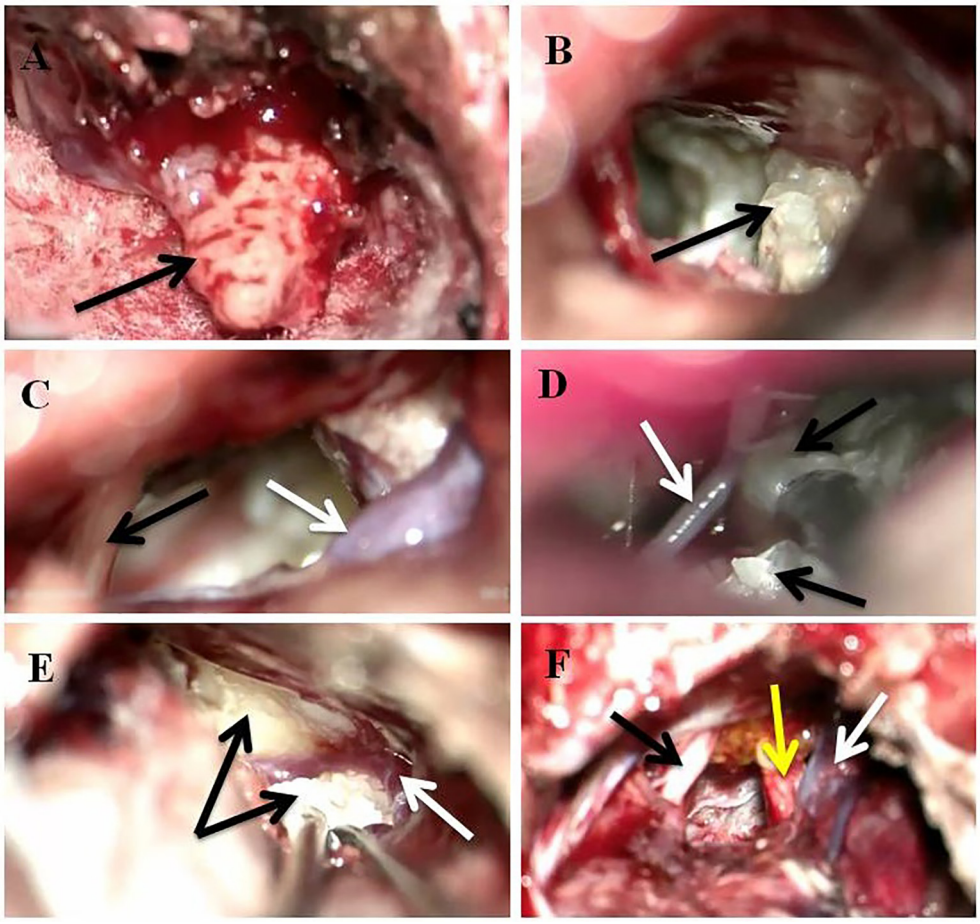

Fig. 2. Intra-operative images. **A.** Initial stage of the operation. Arrow indicates part of subtentorial meningioma node. **B.** Arrow indicates epidermoid cyst (EC) in the cerebellopontine angle (CPA). **C.** White arrow indicates Dandy's vein; black arrow indicates facial-vestibulocochlear nerve complex after removal of EC. **D.** The abducens nerve (white arrow) separated from the EC (black arrows). **E.** The superior cerebellar artery (white arrow) is shown coursing around the EC (black arrows). **F.** Final stage of the operation. Anatomical CPA structures are freed. White arrow indicates Dandy's vein; black arrow, the facial-vestibulocochlear nerve complex; yellow arrow, the trigeminal nerve.

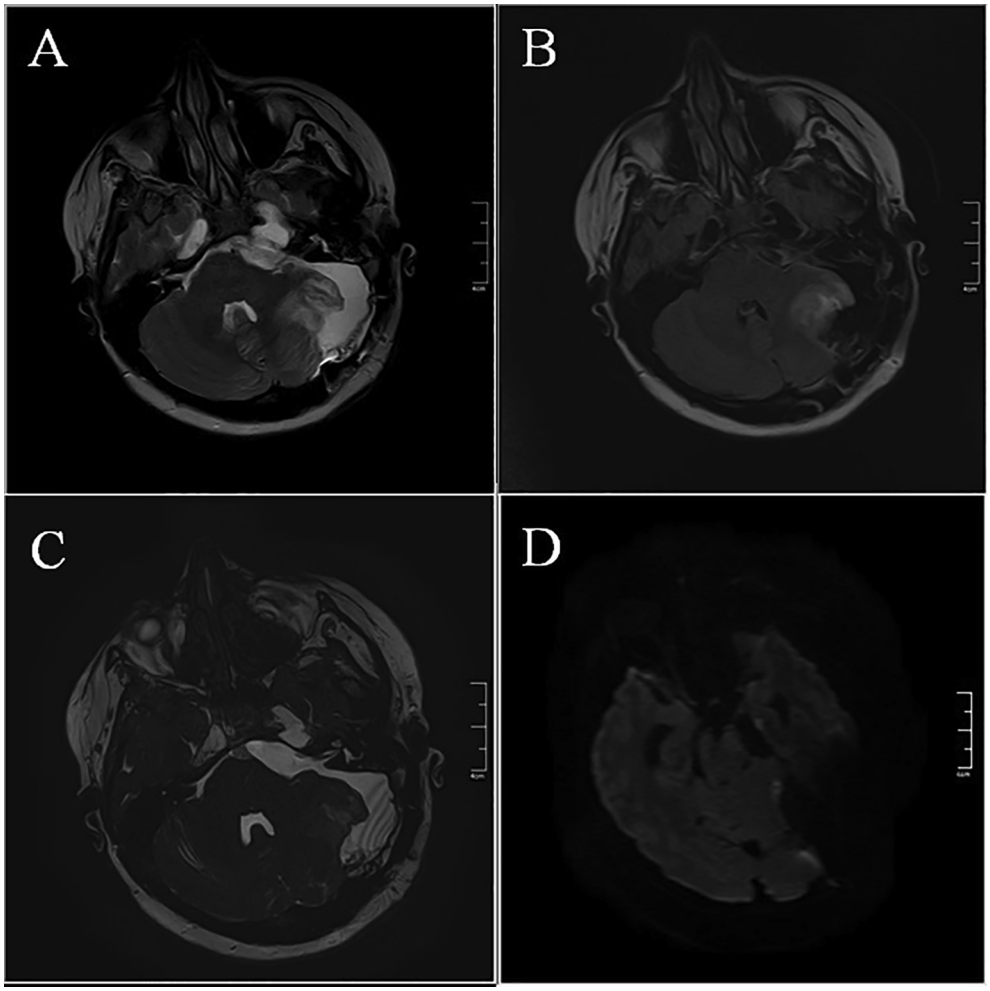

Fig. 3. Post-operative magnetic resonance images (MRIs). **A.** T2-weighted MRI. **B.** Fluid-attenuated inversion recovery image. **C.** = Three-dimensional steady-state free precision image of the internal auditory canal. **D.** Diffusion-weighted image.

(Medtronic, Minneapolis, MN, USA) was performed at all stages of tumour removal. Finally, duraplasty was performed using a pre-prepared fascia lata femoris graft. Pathomorphological response correlated with mixed grade I meningioma and EC. Follow-up CT and MRI findings are depicted in Fig. 3.

Post-operative neurological status included left-sided facial muscles paresis (House–Brackmann grade II), which improved to grade I at 3-month follow-up.

3. Discussion

Meningiomas are common becasue their diagnosis is not particularly difficult. They are usually homogeneous and well-circumscribed, with a broad base adjacent to the dura mater. In approximately 60% of cases diagnosed on CT, meningiomas are slightly hyperdense in comparison with normal brain tissue; in other cases, they are more isodense, and 20%–30% include calcifications. On T1-weighted MRI, 60%–90% appear isointense in comparison with the grey matter, and 10%–40% appear hypointense in comparison with the grey matter. On T2-weighted MRI, 50% are isointense in comparison with the grey matter and approximately 40% are hyperintense. Contrast-enhanced CT and T1-weighted MRI with gadolinium (Gd) usually display intense and homogeneous enhancement [11,12].

The diagnosis of an intracranial EC is quite specific. CT shows the density of ECs to be approximately 0 HU, and they can thus be identical in density to CSF. As in CT, many MRI signal characteristics (e.g. T1 and T2) of ECs are often indistinguishable from those of arachnoid cysts or dilated CSF spaces; however, diffusion-weighted imaging (DWI) and the apparent diffusion coefficient help differentiate them because of the intense amplification of the signal. Contrast-enhanced CT and T1-weighted MRI with Gd do not show enhancement [13].

Due to high occurrence, the pathogenesis of combined glial tumours and meningiomas is best studied. Much attention is given to molecular biomarkers, such as epidermal growth factor receptor (EGFR), vascular epidermal growth factor and p53, which are characterised by similar disruptions of the molecular and signalling pathways [1,10]. Furthermore, meningioma or glioma can induce proliferation in adjacent brain parenchyma or arachnoid cells for tumour proliferation. This can be achieved by increased EGFR expression in tumour cells and evaluated based on the example of meningioma adjacent to glioblastoma [1–3]. Additionally, radiation therapy and surgery play important roles in tumour pathogenesis. Cases of glioblastoma formation in proximity to the radiation or meningioma removal area have been reported [1,3].

Pathogenesis of “tumours by tumour” EC and meningioma has been less frequently studied. To date, there is no convincing evidence of the association or potentiating influence between tumours. Given their relatively rare occurrence, the combination of such tumours is currently considered coincidental. However, Karekezi et al. proposed some interesting theories. The first theory stated that cerebrospinal fluid (CSF) circulating around and under the arachnoid envelope of EC may be an influential factor because it stimulates tumour cell growth via irrigation of the adjacent areas. The second theory was based on EGFR expression in meningiomas and its paracrine stimulation, which may play a role in inducing EC development [10].

According to all available reports, meningioma and EC were located in proximity, indicating a possible local influence on each other. Notably, EC is a heterotopic dysontogenetic tumour originating from epidermal anlage that shifted into the cranial cavity during early embryonic development. This indicates that EC may have formed initially, with subsequent formation of the meningioma. Thus, if the theory of a potentiating influence is true, the tumour most likely originated from EC.

4. Conclusion

A combination of histologically distinct brain tumoursin the same patient, particularly meningioma and EC, is extremely rare. However, the possibility of such a combination should always be considered, and thorough preoperative diagnostics should be performed to prevent complications and misdiagnosis. Currently, co-existence of meningioma and EC is considered coincidental. However, the proximity of these tumours described in previous reports does not exclude the possibility of one tumour's local influence on the other, which should be further investigated.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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