

Brief Report of Special Case

Cavernous malformation of the internal auditory canal

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Summary

Cavernous malformations of the internal auditory canal are a rare clinical entity that, however, should be considered in the differential diagnosis of intracanalicular masses. Even though this type of malformation is usually associated with an evident gadolinium enhancement at MR examination, in some patients, like in this case, the signal characteristics may be not sufficiently specific to allow the correct preoperative diagnosis. Nevertheless, the clinical history, in particular, a rapid onset of cranial nerve deficits, lead to the suspicion of a vascular malformation.

Keywords: Vascular malformation; cranial nerve; intracranial haemorrhage; total removal; auditory canal.

Case report

A 23-year-old woman was admitted to our institution with a 6-month history of progressive hearing loss and a two-week history of right facial palsy, which was initially grade II according to the House and Brackmann scale and worsened acutely (grade IV at admission). A magnetic resonance (MR) study revealed an inhomogeneous right intracanalicular mass extending to the cerebello-pontine angle (CPA). The mass showed a spontaneous high intensity signal that enhanced only slightly after gadolinium administration (Fig. 1A, C). A dilatation of the right internal auditory canal (IAC) was seen on a computed tomography (CT) study (Fig. 1D). Audiometry revealed a hearing loss of 60–70 dB between 1 and 3 Hz on the right side. Cerebral angiography was performed and was negative. The lesion was exposed via a right retrosigmoid approach in the semi-sitting position with neurophysiological monitoring of the facial and the cochlear nerve. The extracanalicular portion of the process visible within the CPA (Fig. 1E) appeared to be mainly a hematoma from recent bleeding. After opening of the IAC the mass could be totally excised with anatomical and functional preservation of the cranial nerves (Fig. 1F). Histologic examination confirmed the diagnosis of cavernous malformation. The post-operative course was uneventful. The facial nerve palsy recovered within the first post-operative days (grade II palsy at discharge) whereas there was no improvement of the hearing deficit.

Discussion

Intrapetrous cavernous malformations involving the facial nerve are a well-known otological entity [8], however only few anecdotic reports of intracanalicular cavernomas (IC) have been published in the literature [3]. IC usually present with hearing loss and/or peripheral facial weakness due to the local mass effect [1, 6, 9]. Differential diagnoses include lesions such as neurinomas, meningiomas, hamartomas, lipomas, cholesteatomas, sarcoidosis, lymphoma, metastasis and Ramsay Hunt syndrome. Involvement of the internal auditory canal in the context of familial and multiple infratentorial angiomas has also been reported recently [1].

MR appearance of extra-axial cavernomas generally shows non-specific findings [5]. Calcification/ossification on CT scans or focal hypointensities on MR images appear more commonly in cavernous malformations than in the other lesions in this location [6]. In our case, dilatation of the right IAC but no calcification was seen on a CT study (Fig. 1D). While IC are reported to have evident gadolinium enhancement [6] our patient showed only a minimal and scattered enhancement (Fig. 1A, C). This phenomenon could be attributed to tissue disruption by the haemorrhage and/or compression by the hematoma. Nevertheless, the suspicion of a vascular pathology was raised because of the characteristics of the clinical history, and in particular, due to the rapid onset of the nerve deficits. The disproportionate character of the symptoms regarding the small size of the lesion on imaging and furthermore the episodic exacerbation of

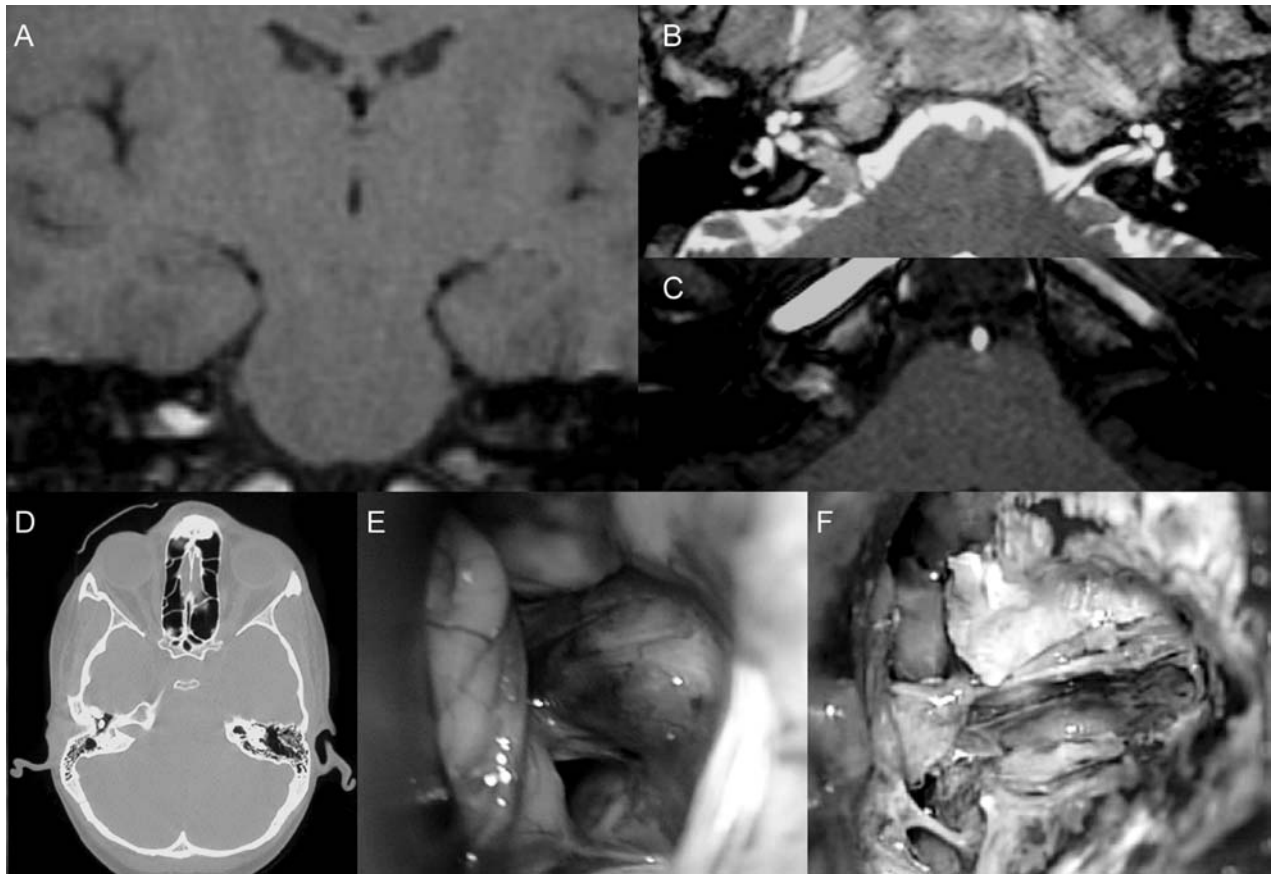


Fig. 1. Coronal (A), and axial (B and C) magnetic resonance imaging showing a right intracanalicular mass that extends minimally to the cerebellopontine angle. The mass showed only slight enhancement after gadolinium administration (A and C). Dilatation of the right IAC but no calcification was seen on a CT bone scan (D). Intraoperative image (E) showing the cavernoma protruding from the internal acoustic canal. The lesion appeared as a reddish soft mass composed by multiple sinusoidal vessels originating from the VIII nerve. The anatomical continuity of the VII nerve was preserved (F)

the symptoms in two stages, probably corresponding to distinct episodes of hemorrhage, were factors suggestive of a vascular lesion.

Facial nerve involvement is commonly found intraoperatively [7]. Pappas *et al.* found the cavernoma attached to the facial nerve in 5 of their 6 cases [7]. The involvement of the nervus intermedius is considerably more rare [10]. Involvement of the inferior vestibular nerve has also been reported previously [4].

On the ground of the relatively slow progression of IC some authors recommend observation and followup [2]. However, repeated bleeding may further injure the cranial nerves leading to their irreversible damage. Moreover, the potential for preservation of the facial nerve function has been reported to be high if surgery is performed early [8]. As illustrated in this case, a prompt surgical treatment in acutely symptomatic patients pro-

vides a chance of complete regression of the clinical symptomatology.

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