Evolution of Vestibular Schwannoma After Removal of Epidermoid Cyst of the Same Location
—Case Report—

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Abstract
A 71-year-old man presented with vestibular schwannoma manifesting as hearing disturbance and truncal ataxia 16 years after removal of a cerebellopontine angle (CPA) epidermoid cyst, and located adjacent to the remnant lesion. The patient first presented with a 6-month history of right trigeminal neuralgia. Neuroimaging demonstrated a right CPA lesion, suggestive of an epidermoid cyst. Right lateral suboccipital craniotomy was performed and the histological diagnosis was epidermoid cyst. A small lesion remained, but the symptoms were relieved. Sixteen years later, the patient presented with right auditory disturbance, vertigo, and truncal ataxia. Magnetic resonance imaging revealed a multiple cystic mass adjacent to the remnant epidermoid cyst in the right CPA. The lesion was removed and the histological diagnosis was vestibular schwannoma associated with the epidermoid cyst. The irritative effect of the remnant epidermoid cyst or surgical procedures may have caused the vestibular schwannoma, but no evidence of the evolution of the different types of tumors was found.

Key words: epidermoid cyst, vestibular schwannoma, collision tumor, multiple brain tumors, evolution tumor

Introduction
Multiple primary brain tumors of different cell types are very rare and account for only 0.3% of all primary brain tumors, excluding cases of phacomatosis or radiation-induced tumors.2,6) Many multiple brain tumors with different cell types have been reported, but most involved coexistent meningioma and glioma.10) There are several hypotheses to explain the association of two different primary intracranial tumors: embryogenic nests causing different brain tumors at different times, systemic carcinogenic factors or metastasis into primary brain tumors, carcinogenic stimulus of adjacent tissues, and growth hormone stimulation, but none have been proved.10,13)

Tumors of the cerebellopontine angle (CPA) account for 10% of intracranial tumors, and are mainly vestibular schwannoma, with 10–15% meningioma, and 2–3% epidermoid cyst.2,6) Collision tumors in this area are rare, with only 15 reported cases: 7 cases of metastatic tumor and vestibular schwannoma, 2 cases of meningioma and vestibular schwannoma, 2 cases of vestibular schwannoma and epidermoid, 1 case of trigeminal schwannoma and epidermoid cyst, 1 case of troclear and abducens nerve schwannoma and meningioma, 1 case of choroid plexus papilloma and vestibular schwannoma, 1 case of epidermoid and glioblastoma, and 1 case of vestibular schwannoma and cholesterol granuloma.1,3–5,7–9,11,12) The coexistence of schwannoma with intracranial epidermoid cyst as a distinct tumor mass has been reported only once.11) Collision tumors of these types forming a single cerebellopontine mass are also rare, with only 2 cases of collision primary tumors.4,5)

We describe a case of collision tumor of epidermoid cyst and vestibular schwannoma on the same side of the CPA, in which vestibular schwannoma was diagnosed 16 years after removal of the epidermoid cyst.

Case Report
A 71-year-old man presented with a 6-month history of right-sided progressive facial pain. On admission, neurological examination disclosed right trigeminal neuralgia. Computed tomography (CT) showed a low density, round mass in the right CPA with a maximum diameter of 1.5 cm. Magnetic resonance (MR) imaging revealed a clearly demarcated lesion, appearing as slightly hyperintense on T1-weighted images, hyperintense on T2-weighted images, and without enhancement after administration of
A right suboccipital craniectomy was performed in the supine position with the head turned laterally. The mass in the right CPA was pearly white, flaky, and avascular, and appeared to be a typical epidermoid cyst. The mass was grossly totally removed by gentle curettage from the trigeminal nerve. The trigeminal nerve and superior cerebellar artery (SCA) were compressed by the tumor and the nerve was directly attached to the SCA. Microvascular decompression was also performed for the trigeminal nerve. No other tumor was observed around the lesion. Histological examination revealed an epidermoid cyst and no malignant findings, and no evidence of other primary tumor (Fig. 2). Postoperatively, the patient made an uneventful recovery and was discharged without neurological deficits. The lesion was almost totally removed. MR imaging showed a small remnant lesion attached to the medulla oblongata but no recurrence was reported (Fig. 3).

Sixteen years after the surgery, he suffered gradual deterioration of right auditory function with diplopia, vertigo, and truncal ataxia. CT revealed an enhanced multiple cystic mass in the right CPA with compression sign to the brainstem. MR imaging showed a new enhanced multiple cystic mass in the internal acoustic meatus on T1-weighted imaging, adjacent to the remnant epidermoid cyst appearing as a hyperintense lesion on diffusion-weighted imaging (Fig. 4). The possibilities of recurrence of the epidermoid cyst or association of another primary tumor were difficult to differentiate. Second removal of the CPA mass was performed through a lateral suboccipital approach. The cystic lesion was an encapsulated solid mass extending throughout the internal acoustic meatus and involving the facial nerve, and appeared to be a typical vestibular schwannoma. Dissection along the facial nerve in this area was difficult, so subtotal removal was performed to preserve the facial nerve. The white pearly lesion of the remnant epidermoid cyst remained adjacent to the rostral side of this cystic mass lesion. The size and appearance of gadolinium (Fig. 1).

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Fig. 4  Magnetic resonance images 16 years after the second surgery revealing a new cystic mass lesion with partially enhanced wall in the right cerebellopontine angle (upper row: T1-weighted images with gadolinium, middle row: T2-weighted images) adjacent to the remnant epidermoid cyst appearing as hyperintense on diffusion-weighted images (lower row).

the remnant lesion had not changed. Histological examination showed vestibular schwannoma without malignant signs associated with epidermoid cyst on the same side of CPA (Fig. 5). His symptoms of vertigo and gait disturbance were relieved after the surgery.

Discussion

The incidence of vestibular schwannoma and epidermoid cyst in the CPA is estimated at one case per 100,000 population yearly. The difference between our present case and the previous two cases was that we could observe the second tumor, vestibular schwannoma. The previous two patients presented with symptoms of 7th and 8th cranial nerve disturbance, and the coexisting tumors were found during surgery. Therefore, which lesion occurred first and how these lesions developed remains unclear. In our case, vestibular schwannoma was not observed at the first surgery for epidermoid cyst in the CPA area. The vestibular schwannoma apparently developed after the occurrence of the epidermoid cyst. Histological examination of the tumor removed at the first surgery found typical epidermoid cyst without malignancy or other cell types. The irritative effect of the remnant epidermoid cyst or surgical procedures may have caused the vestibular schwannoma, but no evidence of the coexistence of the different types of tumors was found. We suggest that the combination of vestibular schwannoma and epidermoid cyst was incidental in our case because no causative mechanism could be identified.

References

3) Gardner WJ, Turner OA: Multiple intracranial tumors: A dis-


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