Concomitant, Contralateral Vestibular Schwannoma and Epidermoid Cyst

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Objective
To report on the coexistence of a vestibular schwannoma and epidermoid cyst on opposite sides.

History & Physical
A 61 year-old woman presented with complaints of severe intermittent headache for 6 months that radiated from the occiput to the frontal region. She had multiple episodes of vertigo lasting 10-15 minutes in duration and 3 episodes of syncope within one month. The patient also reported progressive decreased hearing bilaterally (right greater than left), facial pain in the right V2 distribution of the trigeminal nerve, and bilateral visual changes described as flashes of light. The patient denies tinnitus, otalgia, facial numbness, facial twitching, or loss of facial sensation.

Her otoscopic and general ENT exam were unremarkable, except for tenderness to palpation at the occiput. On tuning fork testing, the Weber lateralized to the left and the Rinne test was positive bilaterally. Her cranial nerve exam was intact.

Figure 1: Normal hearing bilaterally with the exception of a mild sensorineural hearing loss at 2000 Hz in the right ear. Tympanometry revealed normal middle ear pressure and compliance bilaterally.

Results
An audiogram (see Figure 1) was within normal limits with the exception of mild sensorineural hearing loss "notch" at 2000 Hz on the right side, and a similar borderline notch on the left.

A contrast-enhanced MRI (see Figure 2) showed a mass in the left cerebellopontine angle which extended inferior to the left cerebellomedullary junction. This mass predominantly demonstrated cerebrospinal fluid signal intensity on T1 and T2 weighted imaging (see Figures 3 and 4) and showed areas of restricted diffusion weighted images without contrast enhancement, compatible with an epidermoid cyst. It measured approximately 2.5 cm (AP) x 1.0 cm (transverse) x 1.5 cm (cranio-caudal) and likely involved the cisternal portion of the left trigeminal nerve, which was not visualized. Additionally, there was a well defined enhancing mass in the right internal auditory canal near the fundus measuring 4 x 4 x 3 mm, consistent with a vestibular schwannoma.

The patient was advised to obtain an ENG to further evaluate vestibular function. She wished to have the left epidermoid cyst and right vestibular schwannoma observed and followed with serial MRI scans.

Conclusions
Although exceedingly rare, there have been two reported cases of a vestibular schwannoma coexisting with an epidermoid cyst as a single cerebellopontine angle mass.¹ ² Additionally, two cases of contralateral coexisting schwannoma and epidermoid cyst or cholesterol granuloma have been reported.³ However, this represents the first known case of the coexistence of a vestibular schwannoma and an epidermoid cyst on the contralateral side.

Epidermoid tumors are thought to originate in utero and vestibular schwannomas to develop postnatally. Epidermoid cysts are slow growing congenital neoplasms thought to arise from epidermal inclusions. Vestibular schwannomas evolve from the vestibular division of the 8th cranial nerve, with the greatest incidence at 35-55 years.⁴

The simultaneous development of histologically different multiple primary brain tumors, aside from phacomatoses or previous irradiation is very rare. There have been several theories to hypothesize that one tumor may promote the neoplastic transformation of the contiguous cells into different histologic cell types or that a second tumor may be the result of colonization of the neoplastic cellular population.³ These possibilities mostly address the development of adjacent tumors. However, transformation may occur in susceptible areas leading to primary brain tumors with different histologic characteristics and location, which could explain the growth of a primary tumor in a distant site. Further, the existence of systemic carcinogenic principle may also produce systemic tumor disease leading to multiple intracranial neoplasms and other organ tumors.³ The presence of one tumor may act as an irritant, stimulating development of other adjacent tumor or that locally acting oncogenic factors may lead to changes in healthy tissue. Currently, it is unclear if tumors play a role in growth of other tumors.

Epidermoid cysts most commonly spread along the basal surface of the brain, by contagious spread along the normal cleavage lines.³ Arachnoid cysts are known to possess an arachnoidal cap, which is a CSF collection surrounding the tumor.³ Based on this knowledge, perhaps the presence of a vestibular schwannoma and epidermoid cyst could be the result of irritants or oncogenic factors leaking into the CSF leading to the stimulation of tumor growth in susceptible areas. Activation of the receptor for epidermal growth factor (EGF) plays a role in vestibular schwannoma growth. Since ligands for this receptor include EGF and neuregulin (Nrg), it is possible that release of such ligands into the CSF could result in paracrine stimulation of an epithelial rest, leading to formation of an epidermoid cyst.⁵

Figures 2-5 (clockwise from top left):
2 - Post-contrast; 3 - T1 weighted; 4 - T2 weighted; 5 - diffusion weighted.

References