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## Endolymphatic Sac Tumor

**We present** the case of a 48-year-old lady with a history of episodic hearing loss and tinnitus of several years duration. One month prior to consult, there was note of left occipital pain. No history of dizziness, vertigo or facial nerve palsy was elicited. She was neither a smoker nor an alcoholic beverage drinker. No other co-morbidities were elicited. Physical examination revealed a 4-cm diameter left posterior auricular mass which was tender. There was note of a bluish bulge on the left posterior wall of the external auditory canal. The tympanic membrane was intact. The MRI revealed a 5-cm diameter, irregular, avidly enhancing mass at the left mastoid bone with permeative bone destruction and indentation of the left cerebellar hemisphere and left superior temporal lobe but without evidence of brain invasion.

A biopsy was performed followed by a pre-operative tumor embolization then a sub-total petrosectomy with mastoid obliteration. Histologic sections showed an unencapsulated mass with bony invasion composed of cystically dilated glandular structures containing colloid-like material (*Figure 1*) while other areas showed simple and coarse *papillae* (*Figure 2*). The cells were cuboidal to columnar and had a bland cytomorphology with little nuclear pleomorphism (*Figure 3*). Mitoses and necrosis were absent.

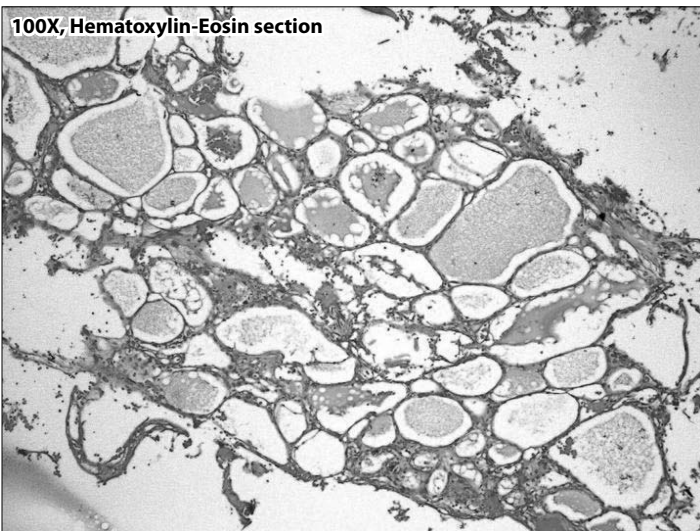
The general histology had a striking resemblance to either normal thyroid tissue or papillary thyroid carcinoma. A TTF-1 immunohistochemical stain however showed negative nuclear staining (*Figure 4*). We signed out the case as an Endolymphatic Sac Tumor. This tumor has been known in the past by such synonyms as “Aggressive Papillary Middle Ear Tumor”, “Heffner Tumor” and “Low-grade Adenocarcinoma of the Middle Ear.” It is rare, affects both sexes in roughly equal frequencies and often presents with hearing and vestibular dysfunctions, facial nerve palsy and a mass. It presents radiologically as a multilocular lytic lesion in the petrous area of the temporal bone with bone destruction.

Because of the histologic resemblance to thyroid tissue, a metastatic thyroid neoplasm is a differential diagnosis. Metastases to this area are rare, cases invariably have a known primary focus and otologic symptoms are uncommon. Immunohistochemical studies

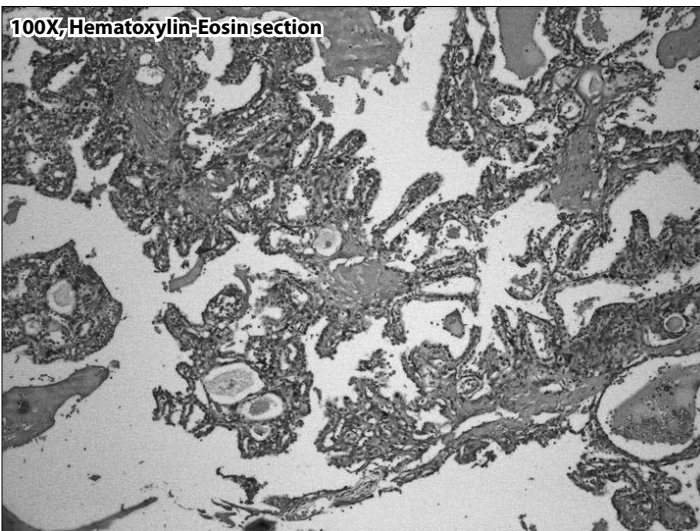
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and clinical correlation are helpful in ruling out a metastasis. Treatment is primarily surgical. Prognosis is generally good but is dependent on the extent of the lesion at presentation. It is locally destructive, has the capacity to damage adjacent nerves and is recurrent if incompletely excised. Death may result from a large, destructive lesion in a vital area.

To date, there are no reports of metastasis which may make the term “adenocarcinoma” not entirely appropriate. We have limited follow-up information on our present case at this time.



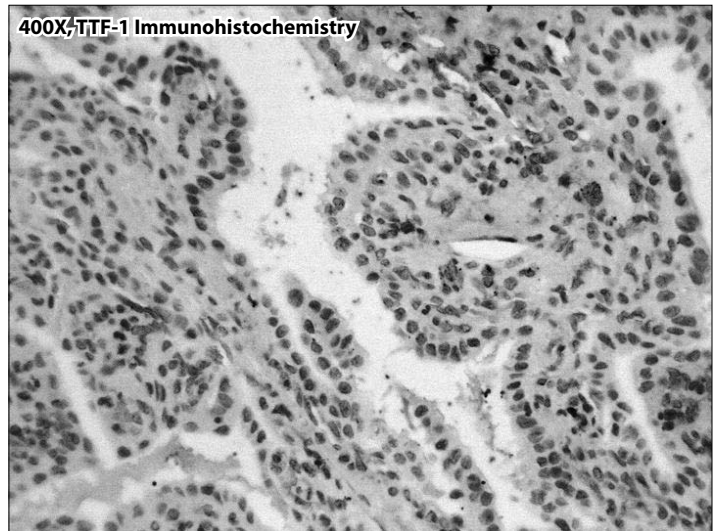
**Figure 1.** Cystically dilated glandular structures containing colloid-like material



**Figure 2.** Simple and coarse papillae



**Figure 3.** Cuboidal to columnar cells with bland cytomorphology, little nuclear pleomorphism and no mitoses or necrosis. Note striking resemblance to normal thyroid tissue or papillary thyroid carcinoma.



**Figure 4.** TTF-1 immunohistochemical stain with negative nuclear staining

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