

Endolymphatic sac tumor

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Endolymphatic sac tumor is an uncommon, locally aggressive tumor. The tumor is located in the medial and posterior petrosal bone region and may involve the dura. A hypervascular tumor involving the endolymphatic sac with destructive changes, it involves the bone and may show reactive new bone formation. Diagnosis is based on clinical, radiological, and pathological correlation. We present a case of endolymphatic sac tumor in a 24-year-old female who presented with a chief complaint of hearing loss.

Case report

A 24 year old female presented with left-sided hearing loss of six months' duration. On examination, the hearing loss was complete. There was mild left-sided facial weakness that became pronounced only on extreme smile. No complaints of vertigo, nausea, or vomiting were present. No features suggestive of Von-Hippel Lindau (VHL) syndrome were present. The patient was further evaluated with high-resolution CT and MRI studies.

High-resolution CT study (Figs. 1 and 2) revealed a 4 x 2.5-cm, expansile, permeative mass lesion along the medial left petrosal bone, with its center in the region of the endolymphatic sac. The mass extended into the left cerebello-pontine angle, internal auditory canal and along the lateral middle ear with involvement of the adjacent mastoid air cells. There was extension of the lesion with erosive changes involving the left bony facial nerve canal in its horizontal segment. The ossicular chain was intact.

On MRI study, the mass lesion was heterogeneous, predominantly hypointense to isointense on T1 with areas of T1 hyperintensity within (Fig. 3).



Figure 1. 24-year-old female with endolymphatic sac tumor. Axial CT image demonstrates a destructive lesion along the medial petrosal bone, in the location of the left endolymphatic sac.

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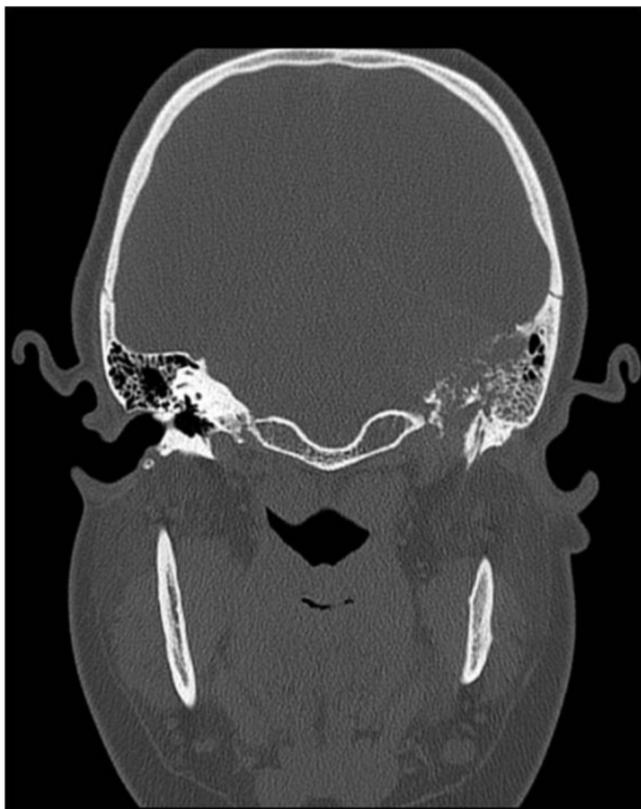


Figure 2. 24-year-old female with endolymphatic sac tumor. Coronal CT image demonstrates erosion of the left petrosal bone and mastoid air cells.

The lesion was hyperintense on T2-weighted sequences with areas of altered gradient susceptibility suggestive of hemorrhage (Fig. 4).

It showed intense postcontrast enhancement suggestive of prominent vascularity (Figs. 5 and 6). Mass effect on the adjacent pons and cerebellum was seen.

The lesion was surgically removed, and histopathology of the resected specimen revealed a locally aggressive tumor (consistent with endolymphatic sac tumor) composed of papillary fronds in simple and complex architecture. The papillae were lined by a single layer of cuboidal and flat epithelium that had moderately clear, foamy cytoplasm. Small foci of hemorrhage were seen. Many dilated vascular channels plus fibrous and hyalinized connective tissue were seen.

Discussion

The endolymphatic sac is an anatomic structure that lies along the posterior and medial petrous temporal bone. The proximal portion is flared and lies in the vestibular aqueduct, covered by bone. The distal portion lies outside the vestibular aqueduct, between the dural layers. The opening of the vestibular aqueduct lies approximately midway between the porus acusticus and the sigmoid sulcus (1, 2). The endolymphatic sac is a network of interconnected si-

nuses and connecting ducts. The middle third has a highly differentiated epithelium of cylindrical cells continuing into irregular crypts and papillae. The subepithelial connective tissue has a rich vascular network (3).

Adenomatous tumors involving the middle ear, mastoid, and petrous bone are broadly categorized into two distinct entities: middle ear adenomatous tumors of nonpapillary

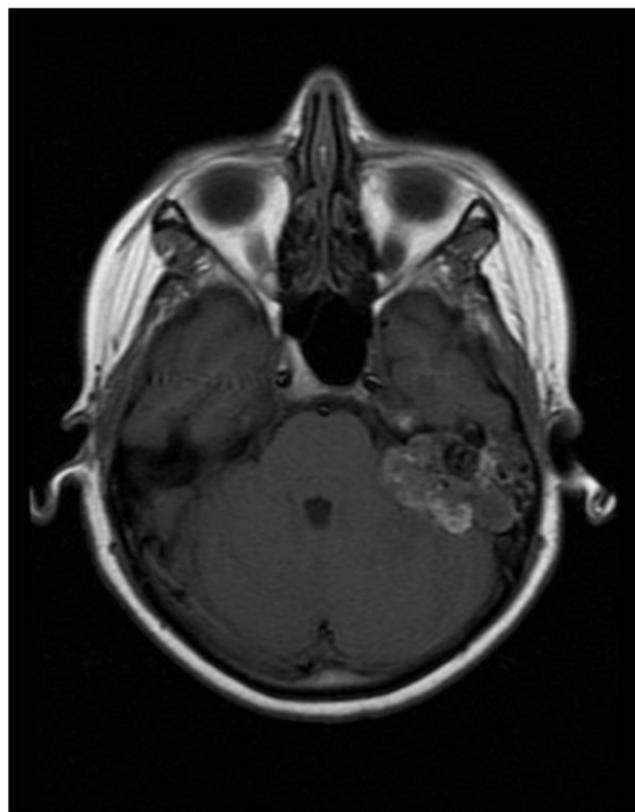


Figure 3. 24-year-old female with endolymphatic sac tumor. Noncontrast-enhanced T1W axial MR image reveals a heterogeneous mass lesion involving the left endolymphatic sac and petrosal bone with extension into the left internal auditory canal. T1 hyperintensities are seen within the lesion on this noncontrast study.

mixed histologic pattern (solid, trabecular and acinar) and adenomas or adenocarcinomas of papillary histologic pattern (4, 5, 6).

Endolymphatic sac tumors are papillary adenomatous tumors. They are slow-growing tumors that may recur locally but do not usually metastasize. They are located in the posterior petrous temporal bone and frequently involve the dura. As the tumor grows, it may involve the supra- and infralabyrinthine, medial mastoid, and posterior tympanic cavity. They are hypervascular and locally invasive and cause destruction of adjacent bone. Reactive new bone formation may be seen. Middle-ear adenomatous tumors are confined to the middle ear and mastoid, are hypovascular, and do not destroy bone.

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Most endolymphatic sac tumors appear to occur sporadically, though an association with VHL disease has been reported in rare cases. In some of these patients with VHL,

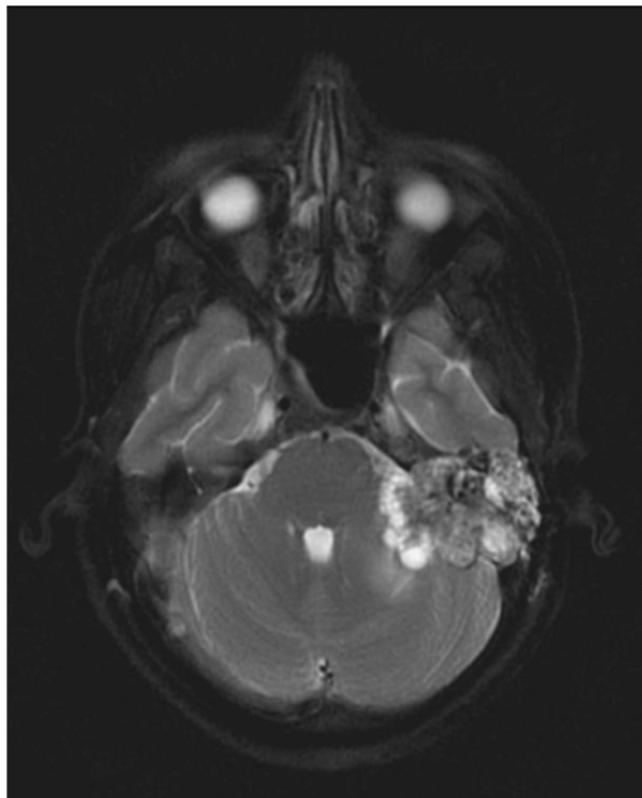


Figure 4. 24-year-old female with endolymphatic sac tumor. T2W axial MR image reveals a heterogeneous mass lesion involving the left endolymphatic sac, petrosal bone, and mastoid air cells.

the endolymphatic sac tumors may be bilateral (7, 8).

Histopathologically, endolymphatic sac tumors generally show papillary adenomatous architecture (9). Papillary fronds in simple and complex architecture may be seen. There may be infiltration into the surrounding connective tissues and bone. Areas of hemorrhage, hemosiderin, and cholesterol clefts may be seen.

The patient generally presents with unilateral hearing loss. Vestibular dysfunction, nausea, and vomiting are other manifestations. Facial-nerve palsy is seen once the tumor becomes large and involves the facial bony canal.

On CT, the tumor is heterogeneous and shows destructive changes involving the adjacent petrous bone, with areas of reactive new bone formation. The tumor bone margins may have a geographic or moth-eaten pattern, and the intratumoral bone may show a spiculated pattern (10). There may be a peripheral rim of calcification, which may represent the expanded cortex of the petrous bone (10). The jugular foramen is generally spared and helps to distinguish the endolymphatic sac tumors from jugulare or

jugulotympanic glomus tumors. The tumor may extend into the internal auditory canal, into the middle ear cavity, and into the region of the bony facial-nerve canal.

On MR imaging, the tumor is generally heterogeneous. On T1-weighted images, the tumor is hypo- to isointense and shows intense postcontrast enhancement. Scattered areas of increased signal intensity on noncontrast, T1-weighted images may be related to the presence of hemorrhage, cholesterol clefts, and proteinaceous cysts in the large

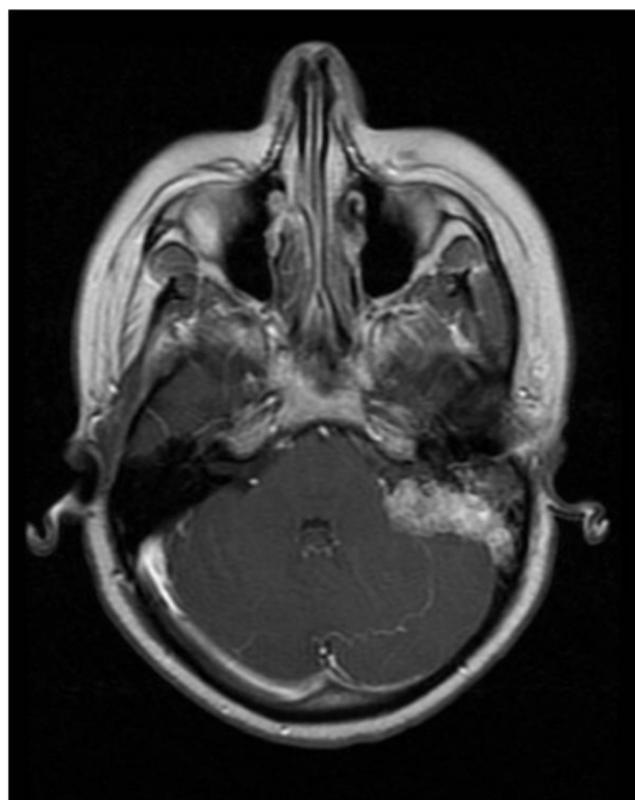


Figure 5. 24-year-old female with endolymphatic sac tumor. Postcontrast T1W axial MR image reveals a heterogeneously enhancing, destructive mass lesion involving the left endolymphatic sac and petrosal bone.

tumors (10). On T2-weighted images, the tumor is heterogeneously hyperintense; on gradient weighted images, it may show areas of altered gradient susceptibility secondary to the foci of hemorrhage.

On angiography, the tumor is highly vascular. The arterial supply is from the branches of the external carotid artery and from anterior inferior cerebellar arteries. The ascending pharyngeal and stylomastoid arteries provide arterial supply in a large number of cases (11). Early venous drainage is not common. Preoperative embolization is helpful and helps surgical excision.

The differential diagnosis would include jugulo-tympanic paraganglioma, vascular tumors like hemangioma, men-

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ingioma, choroid plexus tumors, and bony lesions like chondrosarcomas or eosinophilic granuloma.

Jugulo-tympanic paragangliomas are predominantly infralabyrinthine in location, whereas endolymphatic sac tumors are retrolabyrinthine. The serpentine flow voids of paragangliomas are not typically seen in endolymphatic sac tumors (12).

Intratemporal benign tumors like hemangiomas may have bony spiculations, but they are predominantly in the internal auditory canal, suprageniculate region, or retro-mastoid genu of the facial nerve canal—not in the retrolabyrinthine region (13).

Choroid plexus papillomas involving the foramina of Luschka or cerebellopontine angle are usually intradural but are extraaxial tumors with no bony invasion (14).

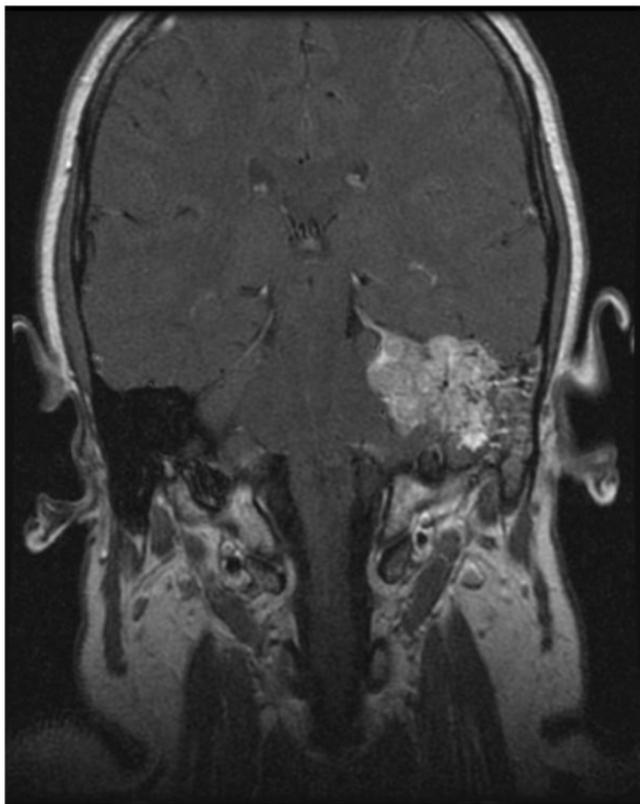


Figure 6. 24-year-old female with endolymphatic sac tumor. Postcontrast T1W coronal MR image reveals a heterogeneously enhancing mass lesion involving the left endolymphatic sac and petrosal bone.

Meningiomas are extraaxial tumors that cause hyperostotic or sclerotic changes and not marked destructive changes (15).

Eosinophilic granulomas generally cause punched-out lytic lesions and involve both the trabecular bone of the mastoid and the dense otic capsule (16).

Chondrosarcoma in the region of the jugular foramen may arise in the petro-occipital fissure and may contain calcific matrix.

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