CASE REPORT

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A Young Case with Endolymphatic Sac Tumors Presented with Right-sided Hearing Loss and Facial Hemi-paralysis

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Abstract

Background: Endolymphatic sac tumors (ELSTs) are uncommon low-grade, locally invasive epithelial tumor that originates from the endolymphatic sac of the inner ear and temporal bone. It is a rare event with less than 300 cases worldwide. In this article, we present a case of ELTS who presented with facial asymmetry and hearing loss. In this article, we present a case of ELTS who presented with facial asymmetry and hearing loss.

Case presentation: A 15-year-old female was admitted to our hospital with right-sided facial hemiplegia and hearing loss in her right ear which was started and progressed within two years. Except for facial hemiparalysis, other physical examinations were normal. In her audiological investigations, severe hearing loss of her right ear was evident. Imaging studies were indicative of ELSTs. This diagnosis was confirmed by histopathological investigations.

Discussion: ELST is an aggressive papillary tumor that arises from the endolymphatic sac. In most cases, this tumor presents as a solitary growth. It usually presents with hearing loss but can be associated with other symptoms like tinnitus, vertigo and nerve VII paralysis. Imaging studies play a crucial role in the diagnosis of ELST, as it can reveal a characteristic soft-tissue mass involving the temporal bone, with bony erosion and extension into adjacent structures. Surgery is considered as the main form of treatment for ELSTs.

Conclusion: ELSTs are exceedingly rare events. Here we presented a case of ESLT with right facial hemiplegia and right ear deafness.

Conflicts of Interest: The Authors declare no conflicts of interest.
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Introduction

Retrolabyrinthine masses are rare, mostly consisting of Endolymphatic sac tumor, paraganglioma, choroid plexus papilloma, and distant metastasis(1). ELSTs are uncommon low-grade, locally invasive epithelial tumor that originates from the endolymphatic sac of the inner ear and temporal bone. It usually invades the semicircular canal, cerebellopontine angle, mastoid bone, posterior petrous bone, and cranial nerve (2).

It is a rare event with less than 300 cases worldwide (3, 4). ELST may be sporadic or associated with von Hippel-Lindau (VHL) syndrome, an autosomal dominant disorder that predisposes to different lesions (5, 6).

Hearing loss and neuropathy based on brain involvement usually manifests ELST. The gold

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standard of ELST treatment is complete surgical excision(7). Here we present a case of ELTS who presented with facial asymmetry and hearing loss.

Case presentation

A 15year-old female was admitted to our hospital with right-sided facial hemiplegia and hearing loss in right ear, which was started and progressed within two years. She did not mention any headache, tinnitus or vertigo. On physical examination the red lesion at posteroinferior of the temporomandibular joint was evident. Furthermore, complete paralysis was found in facial (7^{th}) nerve examination (House-Brackmann grade VI). Otological examination of ear revealed no otorrhea, hyperemia on tympanic drum, ulcer or vesicle. The tympanic drum was normal. He had no nystagmus. Except for facial hemiparalysis, and hearing loss, other neurological examinations were insignificant. No abnormality was observed in her laboratory investigations.

The audiometric examination showed right severe hearing loss. The tympanography of the patient was normal. She underwent spiral temporal bone computed tomography (CT) scan, which revealed "opacification of right mastoid air cells with erosion and destruction septa extension to middle ear, internal ear and external ear with enhancement and also with extra-axial collection. These findings were most consistent with severe acute mastoiditis with complication (Figures 1, 2).

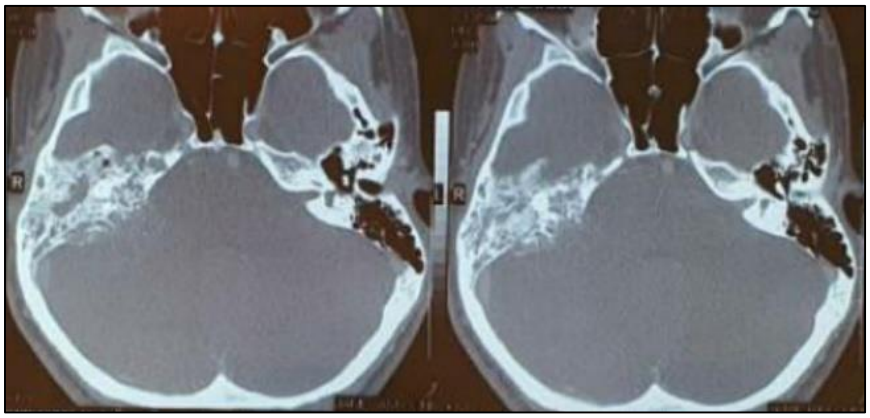

Figure 1. The CT scan of the case.

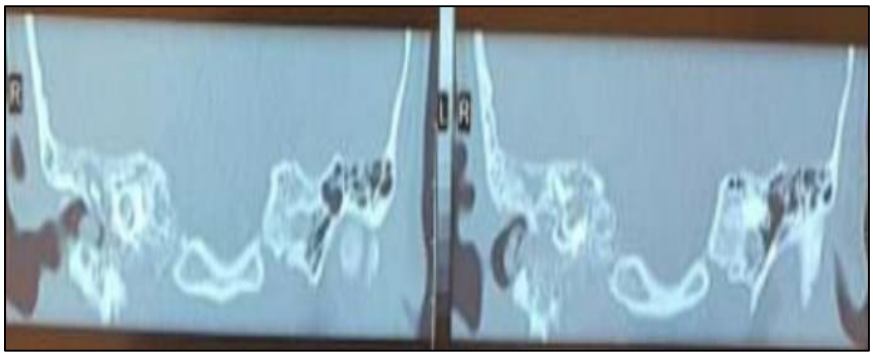

Figure 2. The temporal bone CT scan of the case.

For further investigation, she underwent magnetic resonance imaging (MRI). In MRI investigation, moth eaten or permeative lytic appearance in petrosal bone especially vestibular aqueduct was obvious. A large lobulated lesion with extension to middle ear, jugular bulb, and posterior cranial fossa all indicated ELST.

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The signal of mentioned multilobulated mass is intrinsically T1 and T2 hyperintense, which supports the ELST diagnosis (Figures 3, 4).

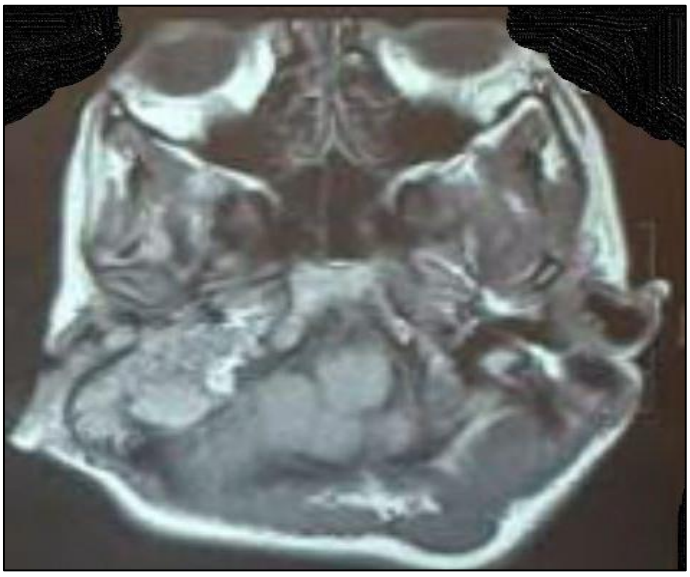

Figure 3. The T1-weighted MRI of the discussed patient.

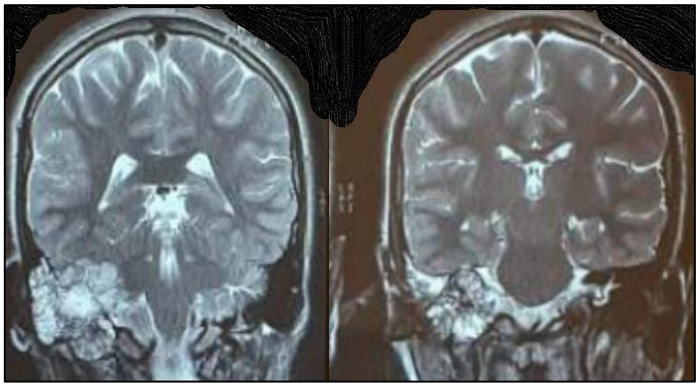

Figure 4. The T2-weighted MRI.

A surgical plan was made for her and the tumor was removed with the cooperation of a neurosurgeon (due to its intracranial component). The tumor was completely excised with transmatoid and translabyrinthine approach. The lesion was sent for pathology. The pathological investigation revealed endolymphatic sac papillary tumor/low grade adenocarcinoma of probable endolymphatic sac origin. On immunohistochemistry (IHC) investigation CK and GFAP were positive. S100 was focally positive. Other evaluated components such as CK, CD10, TTF1, and synaptophysing were all negative.

Early after operation no neurological complication was observed. After six months of follow up no relapse was evident in MRI. Due to her facial paralysis, Trigeminal facial nerve graft was performed successfully.

Discussion

ELST is an aggressive papillary tumor that arises from the endolymphatic sac. In most cases, this tumor presents as a solitary growth, but in around 11-30% of affected individuals, it occurs concurrently with von Hippel-Lindau disease (8). Sporadic types generally appear during the fifth and sixth decades of life. The

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presented case was relatively young in her second decades of life. On the other hand, ELSTs that are linked to von Hippel-Lindau disease tend to occur earlier in life, with a higher frequency in females, and they are bilaterally present around 30% of the time (6).

It usually presents with hearing loss but can be associated with other symptoms like tinnitus, vertigo and nerve VII paralysis. In accordance our case was admitted with hearing loss and facial nerve paralysis, but she stated no tinnitus. Early detection of ELSTs using imaging tests is crucial for preserving hearing in patients with cochleovestibular symptoms(5). Imaging studies play a crucial role in the diagnosis of ELST, as it can reveal a characteristic soft-tissue mass involving the temporal bone, with bony erosion and extension into adjacent structures. In early stages of the ELST destructive lesions in the endolyphatic sac are evident, in MRI investigation the lesion is usually hyperintense (9). The imaging study of the discussed case was also evident of destruction of septa and extension to middle ear, internal ear and external ear. The lesion was hyperintensive in MRI imaging. However, definitive diagnosis requires histopathological examination (10-12).

Early and advances ELSTs differ in the location of involvements. The early stages of ELSTs usually involve sigmoid sinus, medial mastoid bone, and internal acoustic canal. Inferior jugular foramen, posterior fossa dura mater, anterior cavernous sinus, and superior middle cranial fossa, are common affected regions in advanced stages (13).

According to various reports, surgery is considered as the main form of treatment for ELSTs. However, if the aggressive tumors have spread significantly at the time of diagnosis, the chances of a complete cure through surgical intervention are reduced.(14) The use of adjuvant radiotherapy and gamma knife surgery as a treatment method is a topic of debate. Nonetheless, some experts have recommended that postoperative radiotherapy could serve as adjuvant therapy for cases of recurrent disease or partial removal(8). The presented case was treated with complete excision of the mass.

It is recommended that after surgical removal the ELST patients should be followed annually for ten years with imaging studies(15).

Conclusion

In summary, in this manuscript we presented a 15-year-old patient admitted with complete facial paralysis and hearing loss, who was eventually diagnosed with isolated ELST and then successfully treated.

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Conflicts of Interest

The authors declare no conflicts of interest.

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Patients' perspective: Written consent was obtained from the patient. The whole process of examination and the purpose of the article was thoroughly explained.

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