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Letter to the Editor

Endolymphatic sac: A surreptitious anatomy for neurosurgeons

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Dear Editor,

INTRODUCTION

The endolymphatic sac (ES) is an essential structure in the field of otorhinolaryngology, while it can be seen as a hidden corner from neurosurgical perspectives. The ES is a small epithelial-lined channel, considered a non-sensory organ related to the inner ear. The available literature on neurosurgical pathologies and their relation to ES is scattered with a noticeable paucity. In this paper, we attempt to give an overview of the anatomical description, potential functions, and applications of the ES in neurosurgery.

Gudziol and Guntinas-Lichius tapped into the historical journey of the endolymphatic structures, starting from Huscke in 1824 to Hensen in 1880. Their exploration of the Labyrinth, besides its possible connection to the intracranial structures, paved the way to be followed later by Weber-Liel (1823–1891) and their proof of their theory.[9] The theory of existing passable canals between intracranial space and the labyrinth, further in 1986, the exploration of Becker *et al*. related to the drainage of ES presented as a fistulous connection that explained the leakage of the cerebrospinal fluid during ES violating procedures as the stapes surgery.[4,9] Shambaugh GE mentioned that Landquist experimented in 1963 with the installation of colloidal silver into the Scala media of guinea pigs. This resulted in the detection of silver granules only in the ES, thus provoking a debate on the significance and importance of further investigation of the physioanatomy of the ES and duct.[19]

PERTINENT ANATOMY OF ES

The ES represented a segment of the membranous labyrinth, a part of the inner ear composed of two other parts, including the osseous labyrinth and the otic capsule.[6,13] ES has two parts: an intra-osseous (proximal) part medial to the operculum and an intradural (distal) part lying within the two layers of the dura in the endolymphatic fossa near the sigmoid sinus.[14] Lo *et al*. mentioned that the ES is composed of a proximal intraosseous part located within the widening

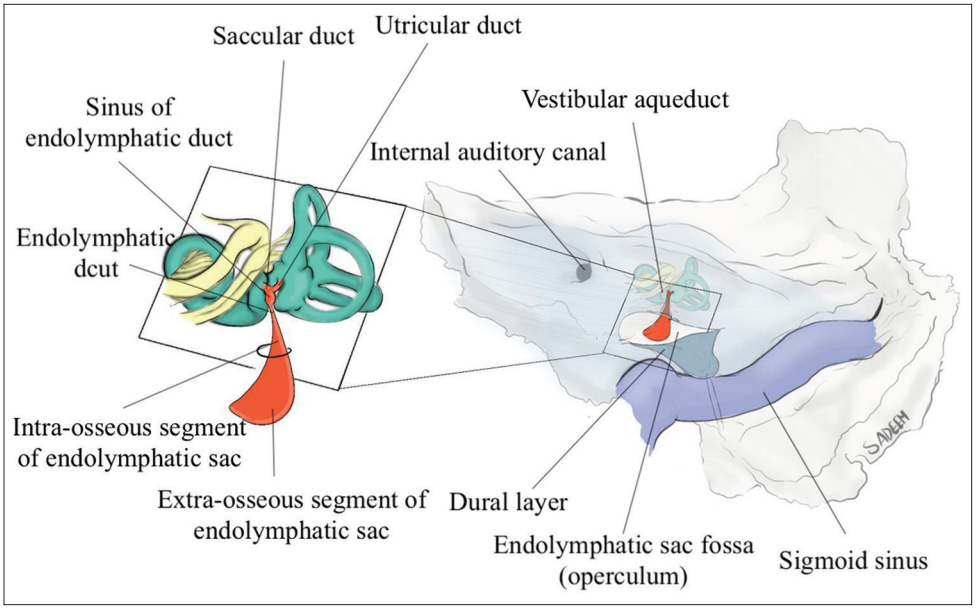

Figure 1: A schematic illustration (the posterior view of the posterior surface of the right petrous temporal bone) detailing the anatomy of the endolymphatic system and its relationship to the surrounding petrous bone; The endolymphatic duct is connected to the membranous labyrinth of the inner ear by the saccular and utricular ducts. The saccular and utricular ducts form the sinus of the endolymphatic duct. The sinus of the endolymphatic duct tapers and becomes the isthmus of the endolymphatic duct as it enters the bony vestibular aqueduct. The isthmus of the endolymphatic duct connects it to the intra-osseous portion (within the vestibular aqueduct) of the endolymphatic duct. Endolymphatic sac tumors arise from the endolymphatic epithelium of the endolymphatic duct with the vestibular aqueduct (osseous portion, striped area); the vestibular aqueduct is considered the site of origin of endolymphatic sac tumors. Distally, the extra-osseous portion of the endolymphatic sac begins as the sac exits the aperture of the vestibular aqueduct. The extra-osseous portion of the sac resides between the leaves of the posterior fossa dura mater on the posterior wall of the petrous ridge.

vertical portion of the vestibular aqueduct (VA) covered on the posterior aspect by thin bone, that is, the operculum. In contrast, the distal extraosseous part sets on the fovea of the posterior wall of the petrous bone with the two layers of the dura.[13] Not more than one-third of the ES lies in the VA of the temporal bone. At the same time, the reminder two-thirds resemble the extra-osseous portion located in the posterior cranial fossa within the duplicator of the dura, as was displayed in Bagger-Sjöbäck *et al*. study.[1] On the 14 dissections of the posterior cranial fossa, the intra-dural portion of the ES was located in a trapezoid thickening of the Dura within the endolymphatic fossa near the operculum.[14]

ANATOMICAL VARIATIONS AND RELATIONS OF ES

Regarding the ES size, variation exists as the mean width of the sac was 12 mm, and the mean length of its superior and inferior borders was 12 mm and 11 mm, respectively.[14] Lo WW *et al*. showed that the intraosseous part varies in size depending on the VA size, which usually varies in width from 3 to 15 mm and 6 to 15 mm.[13] In a study by Friberg U *et al*., the mean width and length of the extraosseous part of the ES were 3.4 and 6.3 mm, respectively.[7]

Highlighting the relation of ES to the sigmoid sinus has been mentioned in different studies, as a study by Lo WW *et al*. showed that in up to 40% of the cases, the distal part of the ES overlaps the sigmoid sinus.[13] Friberg U *et al*. also showed that the ES overlaps the sigmoid sinus in one-third of the cases, and the largest extension distance over the sigmoid sinus is 2 mm.[7] In addition, another study revealed that the ES extended beyond the medial margin of the sigmoid sinus.[14]

Regarding the vasculature of the ES, the arterial blood supply appears to be from the occipital artery.[8] The venous drainage is into the vein close to the distal part of the VA and through the venules directly to the sigmoid sinus.[10]

The function of ES is based on its location. As part of the labyrinth, it serves the purposes of regulating the volume and pressure of the endolymph, modulating the immune response of the inner ear, and eliminating the endolymphatic waste

products by phagocytosis. However, the precise function of the sac is a point of controversy.

PATHOLOGIES RELATED TO THE ES

Lesions related to the ES can vary; an example is the ES tumors which were categorized together with adenomatous tumors of the middle ear and the mastoid air sinus until 1984 when Hassard *et al*. first described an extradural papillary lesion that was adherent to the ES.[11] In 1989, Heffner reviewed the light and electron microscopic and immunohistochemical features of 20 papillary-cystic tumors of the petrous bone and concluded that they were low-grade adenocarcinomas likely of ES origin.[12] Although these tumors do not typically metastasize, a recent case report by Bambakidis *et al*. showed that they could metastasize to distant sites.[2]

There are many lesions affecting the ES reported in the literature. For example, if a tumor is found in the endolymphatic duct, it is considered an extremely rare tumor at the base of the skull. It does not have clear-cut clinical guidelines and can be encountered sporadically from the lymph-epithelium within the vestibular duct, which is characterized by locally aggressive papillary tumors of the petrous bone, which may be sporadic or associated with Von Hippel-Lindau (VHL) disease. However, they can rarely occur in individuals who do not have mutation or deletion of the VHL gene.[15] Their origin is controversial, but as the name suggests, they are thought to arise from ES. Other possible sites of origin include the epithelia of the middle ear and the mastoid air cells.[18]

Radiographic features of ES include the following; generally, ES tumors always arise within the VA [Figure 1], involving the ES or duct. Therefore, the lesion is centered in the posterior (retro-labyrinthine) petrous bone. Regarding computed tomography imaging, it can show erosion of petrous bone in an infiltrative or (moth-eaten) pattern, central calcific speculation, and posterior rim calcification,[17] often intense enhancement. In magnetic resonance imaging, signal characteristics include T1: Most show foci of hyperintensity. T1C + gadolinium: heterogeneous enhancement involving the non-cystic component of the tumor. T2: Heterogeneous signal The tumors affect adults of both genders from 17 to 71 years. The clinical prodrome is prolonged and the most common symptoms include hearing loss, otalgia, tinnitus, vertigo, and facial weakness.[14]

Early detection of these tumors is critical for surgical intervention and may prevent further hearing loss. ES tumors do not metastasize but are highly locally aggressive.[14]

SUMMARY

The ES can be considered an overlooked structure and not fully understood location and function for the general community in neurological surgery. Its location at the junction of the petrous bone and mastoid process renders the ES a potential route for tumors to invade the inner ear canal or spread to the middle and posterior cranial fossa.[3,5,16] In addition, the location of the ES in relation to the sigmoid sinus renders the ES a critical structure in the operative cavity of the related approaches. It can be violated or misinterpreted as another structure if not highlighted in the operative planning.

CONCLUSION

ES is a structure related to the inner ear and can be an asset to the auditory function. It might be an overlooked structure, particularly regarding the value of its pertinent anatomy, variation, pathologies, and operative nuances. The sigmoid sinus renders the ES a critical structure in the operative cavity of the related approaches.

Declaration of patient consent

Patient's consent not required as there are no patients in this study.

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Conflicts of interest

There are no conflicts of interest.

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