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Small papillary tumor in the saccule

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Summary

Papillary tumors of the ear are aggressive neoplasms. Previously, a tumor growing in the saccule had not been reported. We report a tumor found incidentally in the saccule of a patient. Serial sections of both temporal bones of the patient were studied in order to analyze the tumor's origin and influence on audio vestibular function. In the right inner ear, there was a small papillary lesion in the saccule, which looked like a papillary tumor without aggression invasion. The tumor was located in the membranous labyrinth of the saccule, not in the endolymphatic duct and sac. It was not related to Von Hippel-Lindau (VHL) disease, nor was it an endolymphatic sac tumor. The tumor did not influence the hearing and vestibular function in the right ear, although this patient presented a severe sensorineural hearing loss and vestibular function loss because of the vestibular schwannoma in the left ear.

Keywords

tumor; sac of saccule; vestibular function; hearing

1. Introduction

Endolymphatic sac tumors (ELSTs) have only been known as tumor entities since 1984. ELSTs can occur sporadically and are hereditarily connected to von Hippel-Lindau disease (VHL) [1]. This connection was observed in 1992 for the first time and was confirmed by molecular genetic analyses of the VHL gene [2,3]. Papillary tumors of the temporal bone are rare and aggressive neoplasms. ELSTs are aggressive papillary tumors of the temporal bone that are frequently associated with VHL. Convincing anatomic, morphological, and immunohistochemical arguments support an endolymphatic sac origin [4]. Although ELSTs were reported in many patients, a tumor growing in the saccule had not been reported. Here, we describe a tumor growing in the membranous labyrinth of the saccule, and analyze its origin and influence on vestibular function.

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2. Materials and methods

Ethical approval and consent were obtained for the removal of the tissue for scientific purposes. The temporal bones of the patient were removed during autopsy about half an hour after death. They were fixed in 10% formaldehyde, decalcified in 10% EDTA, dehydrated and embedded in celloidin according to routine methods, and serially sectioned horizontally or vertically at 20 μm . Every 10th section was stained with hematoxylin-eosin (H&E), observed with light microscopic examination (Nikon Microphot-FXT), and analyzed according to the clinical and historical data.

3. Results

3.1 Clinical course

The patient was a 74 year old woman who presented left hearing impairment. She suffered from left breast cancer and underwent mastectomy of the left breast at 58 years old. She complained of left hearing impairment and visited the clinic at 71 years old. Examination of pure tone auditory function showed sensorineural hearing loss in the left ear, but normal function in the right ear. Auditory brainstem response (ABR) at 100 dB HL showed no response in the left ear and a normal response in the right ear. Vestibular function using the Caloric test with ice water showed no response in the left ear and a normal response in the right ear. Electronystagmography (ENG) showed no spontaneous nystagmus, no positional nystagmus, and no gaze nystagmus. Radiological examination using MRI showed a medium-sized tumor in the cerebellopontine angle and in the internal auditory canal of the left ear, and did not showed any tumor in the right ear. The diagnosis was vestibular schwannoma in the cerebellopontine angle and in the internal auditory canal of of the left ear. She died of sepsis at 74 years old. The autopsy found metastatic cancers in the right upper lobe of the lung, the right temporal lobe of the brain, and the left cerebellopontine angle.

3.2. Temporal bone findings

In the right temporal bone, the number of vestibular ganglion cells was normal; some had a yellow cytoplasmic pigment like plaque. Superior and inferior vestibular nerves, the cochlear nerve, and the facial nerve were well preserved and the vestibular organs were normal. There was a small papillary lesion in the saccule (Fig. 1). The lesion projected from the proximal part of the saccula membrane and was connected by a narrow neck with the saccula membrane. The base of the tumor was lined by cuboidal epithelium. The stroma of the tumor appeared to be dense collagen. The tumor adhered to the macula and grew to other sides so that the structure of the maculae of the saccule was normal (Fig. 2). The tumor was located in the membranous labyrinth of the saccule, not in the right endolymphatic duct and sac. It was not near the orifice of the right endolymphatic duct. The right endolymphatic duct was not enlarged. The utricle and the semicircular canals were intact. The structures of the cochlear, middle ear were normal (Fig. 3).

In the left temporal bone, the diameter of the internal auditory canal was enlarged to approximately twice its normal size and a vestibular schwannoma occupied most of the canal. The cochlear nerve and lower vestibular nerve were replaced by the schwannoma. Destruction of vestibular and cochlear nerve fibers was noted in the internal auditory canal.

2. Discussion

Papillary tumors of the temporal bone are aggressive neoplasms, which may occur sporadically or as a part of von Hippel-Lindau disease. An aggressive papillary middle ear/temporal bone tumor (APMET) and a benign papillary tumor of probable mesonephric origin (APMO) may

represent major visceral manifestations of VHL [5]. Tumors of the endolymphatic sac identify the origin of these rare tumors. The clinical manifestations are sensorineural hearing loss, facial paralysis, cerebellar disorders and vertigo. These tumors are locally invasive, destructive, hypervascular, and exhibit consistent imaging and histopathologic features [6]. These aggressive papillary tumors typically invade the mastoid bone and otic capsule, and can grow into the petrous apex [7,8]. They classically grow very slowly, resulting in late clinical manifestations with an expansive mass invading the temporal bone and extending into the posterior fossa. Radiologically, these endolymphatic sac tumors can mimic metastatic carcinoma, paraganglioma, or cerebellar haemangioblastoma, especially in von Hippel-Lindau disease. Histology shows a papillary epithelial tumor with hypervascular stroma. In one study, retrolabyrinthine bone destruction was centered at the external aperture of the vestibular aqueduct in all four patients with ELSTs who underwent computed tomography (CT) and magnetic resonance (MR). These radiological changes may help distinguish ELSTs from other tumors of the temporal bone and posterior fossa [9]. In our observation, the tumor in the saccule looked like a papillary tumor. When the tumor attached to the maculae of the saccule, it grew to other sides so that the structure of the maculae of the saccule was intact. It only grew in the saccule, with no evidence of aggression to any other tissue, possibly because it was so small. Blood vessels were not clear in the tumor, possibly because the tumor was well fed from the endolymph. All the symptoms of VHL and ELSTs were not presented in this patient.

Tumors of the endolymphatic sac are locally invasive neoplasms arising in the temporal bone that can cause hearing loss, tinnitus, vertigo, aural fullness, and facial-nerve dysfunction [10]. ELSTs, which often are associated with von Hippel-Lindau (VHL) disease, cause irreversible hearing loss and vestibulopathy. A study showed that audiovestibular symptoms were presented in three patients before a tumor was evident on neuroimaging. Successful resection of the ELSTs was accomplished by performing a retrolabyrinthine posterior petrosectomy (RLPP). Hearing stabilized and vestibular symptoms resolved after surgery in all patients [11]. In our observation, the tumor grew on the right side, but the hearing and vestibular functions were normal on the right. Perhaps the tumor was so small that it did not influence them. The saccule maybe supplied a space and permitted some changes in it without influencing the vestibular function. Audio-vestibular symptoms were the most common symptoms present in patients with acoustic neuroma [12]. Therefore, this patient presented a severe sensorineural hearing loss and vestibular function loss on the left side.

Von Hippel-Lindau (VHL) disease is an uncommon, autosomal dominant hereditary multitumor syndrome caused by germline alterations of the VHL gene, which has been mapped to chromosome 3p25–26, and recently identified as a tumor suppressor gene [13]. The major lesions in VHL disease include hemangioblastomas in the central nervous system, retinal and renal cell carcinomas, pheochromocytomas, pancreatic tumors, epididymal cystadenomas, endolymphatic sac tumors, carcinoid tumors, and multiple cysts of the kidney, pancreas, and epididymis. Histologic features of VHL tumors include a high degree of vascularization and the presence of a clear cell component [14]. ELSTs are rare intracranial tumors originating from the pars rugosa of the endolymphatic sac [15]. The patient in our study presented left breast cancer, vestibular schwannoma in the left ear, metastatic cancer in the right upper lobe of the right lung, metastatic cancer in the right temporal lobe of the brain and metastatic cancer in the left cerebellopontine angle. The tumor in the saccule grew in endolymphatic liquid and originated from the saccule membrane. It was the only tumor found in the left ear; no other tumors were found anywhere in the body. This patient was 74 years old and demonstrated no symptoms of VHL disease. In summary, this tumor in the sac of the saccule was neither related to Von Hippel-Lindau (VHL) disease, nor belonged to the endolymphatic sac tumor class. Further details about this tumor, such as the natural history and anatomical origin, need to be further researched.

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Fig. 1. A small, cauliflower-like tumor with a papilla in the the saccule
Scale bar = 100μm.

Fig. 2.

This tumor is connected by a narrow neck with the saccule membrane and adheres to the macula. The base of the tumor is lined by a cuboidal epithelium. The stroma of the tumor appears to consist of dense collagen. Scale bar =50 μ m.

Fig. 3.

A small tumor was located at the membranous labyrinth in the right saccule and touched the saccular macula (**ms**), not situated in the right endolymphatic duct (**ed**) and sac. The right endolymphatic duct was not enlarged. The utricle and the posterior semicircular canal (**psc**) were intact. The structure of the cochlear, middle ear was normal. Scale bar =200 μ m