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CASE REPORT

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 neither 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.

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.

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,

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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 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).

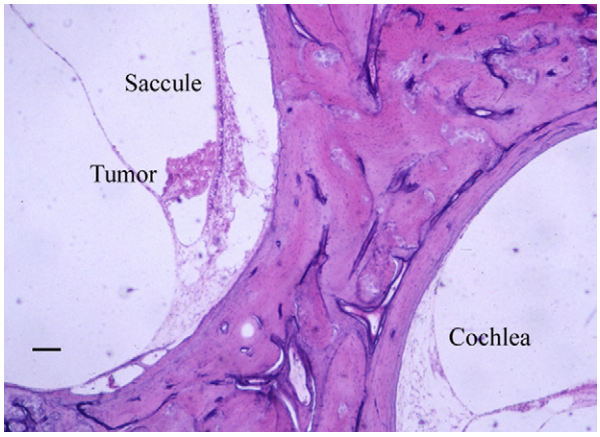

Fig. 1 A small, cauliflower-like tumor with a papilla in the saccule. Scale bar = 100 μm.

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

4. Discussion

Papillary tumors of the temporal bone are aggressive neoplasms, which may occur sporadically or as a

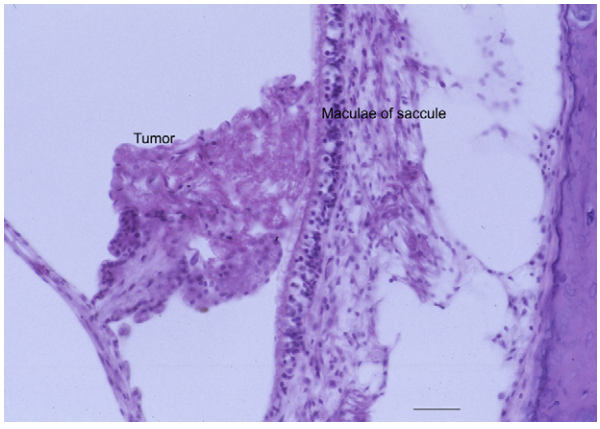

Fig. 2 This tumor is connected by a narrow neck with the saccula membrane and adheres to the macula. The base of the tumor is lined by a cuboidal epithelium. The stroma of the tumor appears to consist of dense collagen. Scale bar = 50 μm.