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Neuroendocrine tumor of the middle ear

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A 39-year-old female patient was referred by a laryngologist to our center due to conductive hearing loss for further diagnostics, with no other abnormalities in the laryngological examination. Laboratory tests showed no abnormalities. The magnetic resonance imaging performed in the middle ear on the left side, at the promontory level, revealed the presence of an irregular focal lesion, which was intensely contrast-enhanced, with features of diffusion limitation (Figure 1A). The lesion extended to the eustachian tube, with the closure of its opening, accompanied by massive secondary effusion within the cells of the mastoid process (Figure 1B). CT examination excluded bone destruction of the scutum, auditory ossicles and bony labyrinth (Figure 1C).

Due to the suspicion of a malignancy, the patient was referred for an urgent operation. The tumor of the tympanic cavity and mastoid process on the left side was removed. Histopathological examination revealed a tumor composed of a homogeneous population of

cells forming rosettes, single solid fields and tubules with mucus (mucicarmine+) presence (Figure 1D). Angioinvasion and perineural infiltration were not detected. Cancer cells expressed synaptophysin, chromogranin, CD56, CK AE 1/ AE 3, and focally CK7. (Figure 1E). The tumor presented low mitotic activity: mitotic index was 1/10 HPF, ki67 2%. According to the WHO classification of neuroendocrine tumors from 2019, the tumor was classified as middle ear neuroendocrine tumor G1 (MeNET G1). Due to diagnostic difficulties, the material was consulted in a second center, where the diagnosis was confirmed. No signs of distant spread were detected in the chest, abdominal cavity and pelvis CT scan. The patient remains under follow-up in our clinic.

Neuroendocrine tumors (NENs) are rare cancers that mainly develop in the digestive system and lungs. Middle ear NENs constitute less than 2% of primary ear tumors [1]. The most common symptoms experienced by patients are unilateral hearing loss and tinnitus, less frequently, pain, discharge in the ear or facial nerve palsy [1, 2]. The middle ear NEN usually surrounds the ossicular chain without destroying it. Perforation of the eardrum with the spread of the tumor to the ear canal occurs sporadically, as does the destruction of the surrounding bone [3].

Immunohistochemical staining for chromogranin A, synaptophysin and CD56 is necessary to confirm the histopathological diagnosis. The Ki67 proliferation and mitotic indexes are necessary for grading neuroendocrine tumors. Hence, they are essential in prognosis and therapeutic decisions [3, 4].

In the available literature, cholesteatoma was often the first suspicion. In our case, it was a paraganglioma, which is taken into account in the differential diagnosis due to its very strong vascularization [2]. The studies describe middle ear NEN as a low-grade malignant lesion with rare local recurrences and distant metastases [5].

The primary treatment method is complete surgical resection. Adjuvant treatment is not recommended. Some authors propose that radiotherapy and chemotherapy may be used as postoperative adjuvant therapy when complete resection is not possible. After surgery, long-term follow-up is recommended due to slow progression and difficulties in resection with wide margins [5].

Article information

Conflict of interest None declared.

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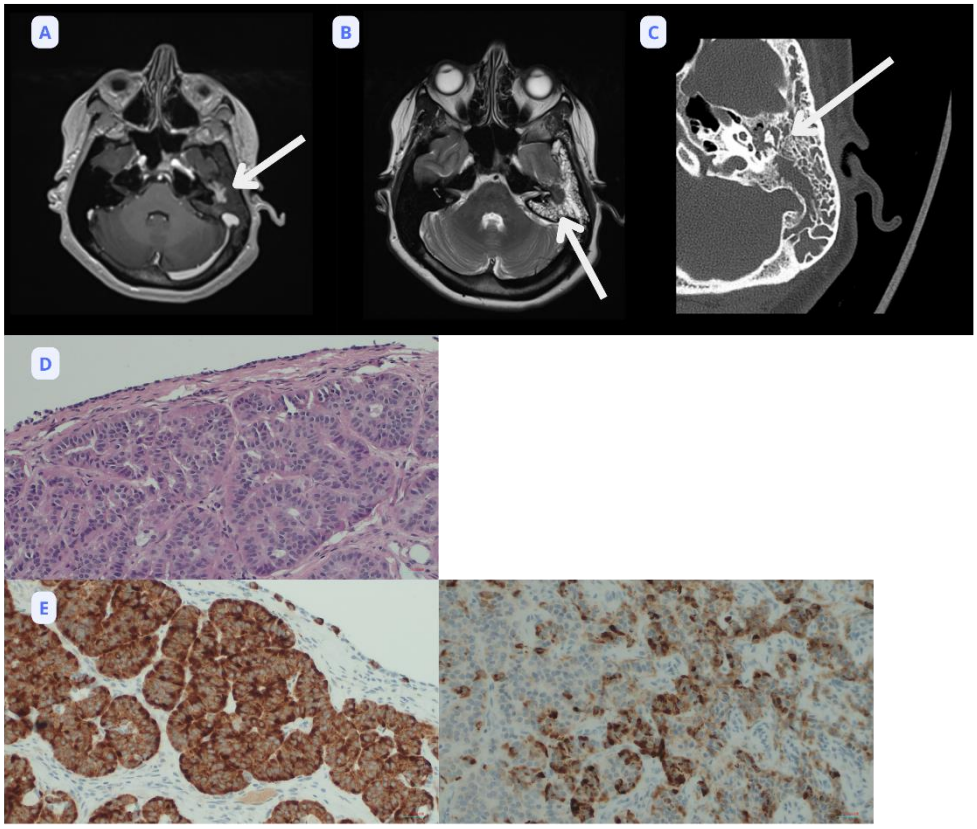

Figure 1 A – Magnetic resonance imaging scan (T1+C) showing a lesion in the middle ear with intense contrast enhancement; B – magnetic resonance imaging scan (T2) showing massive effusion within the mastoid cells; C – the temporal bone computed tomography showing a lesion in the middle ear without ossicular chain erosion; D - histopathological examinations showing infiltrate composed of monomorphic cells forming rosette and nest-like structures. (hematoxylin-eosin staining, magnification ×200); E - immunohistochemical examinations: on the left synaptophysin +, on the right chromogranin +