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Middle ear tumors rather than inflammatory and infective lesion: retrospective histopathological evaluation of eleven cases

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Abstract

Background: Middle ear tumors are a rare group of neoplasia due to their location. In this study, we analyzed our archives of the Department of Pathology for the middle ear tumor cases diagnosed between 2010 and 2019.

Results: Within the mentioned period, 591 cases were operated on for middle ear tumors. Eleven of these cases were diagnosed as middle ear tumors other than cholesteatoma. Considering the distribution of the cases according to histopathological diagnoses, 5 (45.4%) paragangliomas, 4 (36.3%) schwannomas, and 2 (18.2%) meningiomas were detected. No recurrence was observed in the clinical follow-up of the patients after excision.

Conclusions: Although tumors located in the middle ear, which generally cause similar clinical complaints, are rarely seen, the tissue should be evaluated histopathologically because it will affect the treatment and prognosis of the patients.

Keywords: Middle ear, Paraganglioma, Meningioma, Schwannoma

Background

Ear tumors were previously divided into three groups: the outer ear, middle ear, and inner ear tumors according to their location. In the World Health Organization (WHO) 2017 Head and Neck Tumors latest classification, tumors located in the inner and middle ear are mentioned under a common heading, since the region where the tumor originated cannot always be determined [1].

Middle ear tumors constitute a rarely encountered neoplasia group [2]. This group includes schwannoma, meningioma, middle ear adenoma, paraganglioma, endolymphatic sac tumor, aggressive papillary tumor, and squamous cell carcinoma, according to the WHO Head and Neck Tumors latest classification. Although cholesteatoma is a non-neoplastic lesion, it is included in the tumor classification due to its ability to destroy local structures and show recurrence [1].

In this study, middle ear tumors other than cholesteatoma diagnosed in our hospital were retrospectively analyzed, and their clinical and pathological features were mentioned.

Methods

In this study, we analyzed archives of the Department of Pathology for the middle ear tumors cases diagnosed between 2010 and 2019. The hematoxylin-eosin-stained slides were reexamined. The histopathological features were documented. The cases were evaluated retrospectively according to age, gender, tumor localization, clinicopathological features, and prognosis. Our study was approved by the local Clinical Research Ethics Committee dated May 13, 2020, and numbered 2020-8/28.

Results

Within the mentioned period, 591 cases were operated on for middle ear tumors. Eleven of these cases were diagnosed as middle ear tumors other than cholesteatoma. The clinical information of the cases is given

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in Table 1. Eight (72.7%) of the cases were females and 3 (27.2%) were males. The age distribution ranged from 25 to 70 years, with a mean age of 49. Six (54.5%) of the cases were located in the right ear and 5 (45.4%) in the left ear. The most common complaint was found to be hearing loss. Considering the distribution of the cases according to histopathological diagnoses, 5 (45.4%) paragangliomas, 4 (36.3%) schwannomas, and 2 (18.2%) meningiomas were detected. No recurrence was observed in the clinical follow-up of the patients after excision.

Of the 5 paraganglioma cases, 1 was male (20%) and 4 were females (80%). The mean age of these cases was 51.2 years (range: 25–70 years). Three (60%) of the cases were located in the left ear and 2 (40%) in the right ear. In the microscopic examination, it was observed that tumor cells form nests, especially in the perivascular areas. Some of the tumor cells were epithelioid with large eosinophilic cytoplasm while some were spindle-shaped. No atypia, mitosis, or necrosis was observed. Tumor cells exhibited strong immunopositivity with neuroendocrine markers such as synaptophysin and chromogranin, CD56. The ki-67 proliferation index of the tumor was found around 0.05% (Fig. 1).

Of the 4 schwannoma cases, 2 were males (50%) and 2 were females (50%). The mean age of these cases was 45.5 years (range: 43–49 years). One (25%) of the cases

Table 1 Patient demographics, pathological diagnosis, treatment, and disease status of cases with middle ear tumours

Case	Age	Sex	Tumour location	Pathological diagnosis	Treatment	Prognosis
1	44	F	R	Schwannoma	Excision	No evidence of disease
2	45	M	R	Paraganglioma	Excision	No evidence of disease
3	48	F	L	Paraganglioma	Excision	No evidence of disease
4	70	F	L	Paraganglioma	Excision	No evidence of disease
5	64	F	R	Paraganglioma	Excision	No evidence of disease
6	25	F	L	Paraganglioma	Excision	No evidence of disease
7	49	F	L	Meningioma	Excision	No evidence of disease
8	56	F	R	Meningioma	Excision	No evidence of disease
9	46	F	L	Schwannoma	Excision	No evidence of disease
10	43	M	R	Schwannoma	Excision	No evidence of disease
11	49	M	R	Schwannoma	Excision	No evidence of disease

M male, *F* female, *L* left, R:right

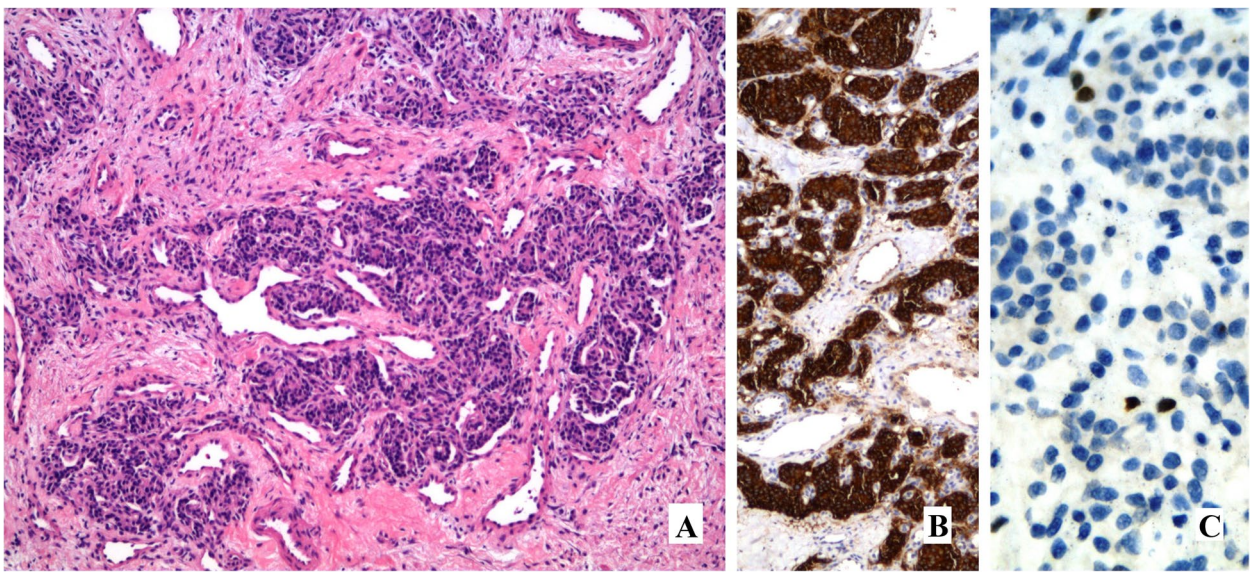

Fig. 1 Photomicrographs of the middle ear paraganglioma. **A** Tumor cells consisting of epithelioid and spindle cells showing nests in perivascular areas (HE ×100). **B** Positivity of diffuse strong synaptophysin in tumor cells (IHC ×100). **C** Ki-67 proliferative index in tumor cells (IHC ×400)

had a tumor in the left ear and 3 (75%) in the right ear. Microscopic examination revealed a tumoral lesion consisting of a tight arrangement of cells with curled nuclei and fibrillar cytoplasm. No atypia, mitosis, or necrosis was observed. Tumor cells showed diffuse positive staining with S-100 immunohistochemical stain (Fig. 2).

Both of the 2 meningioma cases were females (100%). The mean age of these cases was 52.5 years (range: 49–56 years). One (50%) of the cases were located in the left ear and the other one (50%) in the right ear. In the microscopic examination, it was observed that the syncytial tumor cells had indistinct cell membranes and eosinophilic cytoplasm that exhibited a whorl-like structure. Tumor cells showed diffuse staining with epithelial membrane antigen (EMA) and progesterone receptor (PR) immunohistochemical stains.

Discussion

Middle ear tumors originate from structures (bone tissue, epithelial, nerve tissue, etc.) in the middle ear [3, 4]. Among these tumors are schwannoma, meningioma, middle ear adenoma (benign neoplasia), squamous cell carcinoma, endolymphatic sac tumor, and paraganglioma (malign neoplasia). The aggressive papillary tumor is also included in local invasive neoplasia [1].

The most common tumors in the middle ear are schwannoma (40%), paraganglioma (15%), middle ear adenoma (9%), and squamous cell carcinoma (5%) [5]. According to some sources, paraganglioma is the most common, followed by schwannoma and congenital cholesteatoma [6, 7]. In our series, paraganglioma was the most common with a rate of 45.4%.

The paragangliomas are also known as glomus tumors or chemodectomas. Paragangliomas are frequently seen in women (approximately 66–90%). It can be encountered in a wide age range (often around 55 years old). It shows a bimodal peak incidence in the 4th and 7th decades of life. It can cause symptoms such as hearing loss, tinnitus, dizziness, facial nerve palsy, and pain. It may accompany familial syndromes (von Hippel Lindau, neurofibromatosis type 1, succinate dehydrogenase mutations, etc.) such as paragangliomas seen in other regions. Most middle ear paragangliomas are small in size (approximately 2–3 mm). Magnetic resonance imaging (MRI) helps in recognizing the characteristic vascularity of the tumor [8]. Microscopically, epithelioid or spindle-shaped cells are seen to form nests or layers, and stroma has a rich vascular network. Immunohistochemically, staining is observed with neuroendocrine markers such as Neuron Specific Enolase (NSE), synaptophysin, and chromogranin, CD56. Sustentacular cells show positive immune staining with S-100 [5, 9]. The clinical and morphological findings of our paraganglioma cases were found to be compatible with the literature data.

Schwannomas are generally seen over the age of 40. It shows unilateral localization. However, it is observed bilaterally and at younger ages in cases with a syndrome such as neurofibromatosis type 2. They are generally encapsulated well-circumscribed lesions. As in schwannomas seen elsewhere, it consists of Antoni A and B

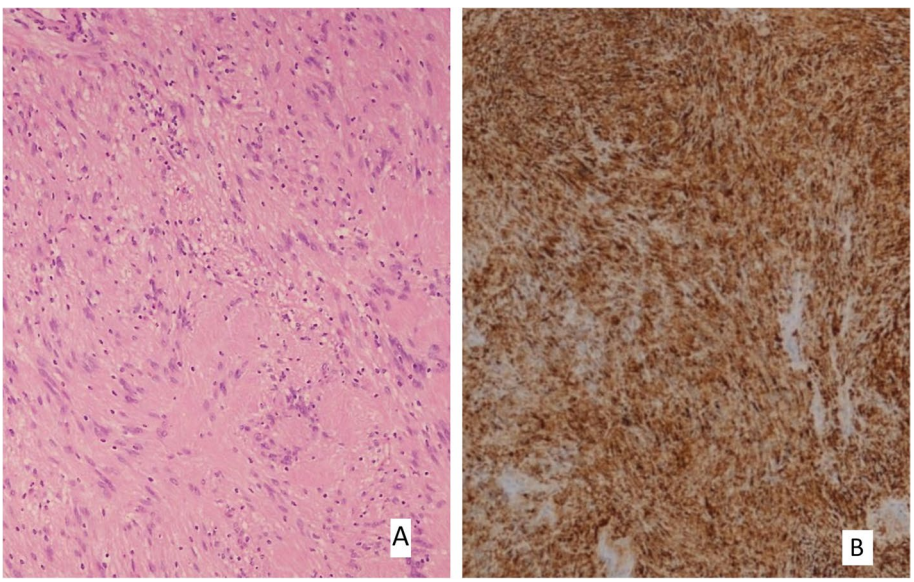

Fig. 2 Photomicrographs of schwannoma. **A** The tumor consists of a tight arrangement of cells with curled nuclei and fibrillar cytoplasm (HE ×100). **B** Positivity of diffuse S-100 in tumor cells (IHC ×100)

fields and Verocay bodies. Hyalinized vessel walls are encountered. Degenerative changes such as hemorrhage, necrosis, and myxoid changes can be observed. Immunohistochemically, they show a positive immune reaction with S-100 [5, 10]. The clinical and morphological findings of our schwannoma cases were found to be compatible with the literature data.

Meningiomas are more common in women. The mean age at diagnosis is 50. It causes complaints such as hearing loss, headache, dizziness, and tinnitus. These symptoms are often long-lasting. Histologically, cells with round or oval nuclei with indeterminate cell borders form nests. Intranuclear pseudo-inclusions and psammoma bodies are frequently seen. Bone destruction is seen in 1/3 of the cases. Immunohistochemically, tumor cells show positive staining with EMA and PR [4]. The clinical and morphological findings of our meningioma cases were found to be compatible with the literature data.

There was no aggressive papillary tumor, endolymphatic sac tumor, squamous cell carcinoma, and middle ear adenoma in our case series.

When the literature is reviewed, there are lots of case reports of a wide variety of tumors located in the middle ear, such as ameloblastoma, teratoma, capillary hemangioma, inverted papilloma, lipoma, myxoma, odontoma, hemangioendothelioma, melanoma, synovial sarcoma, plasmacytoma, carcinosarcoma, primary intestinal-type adenocarcinoma, and rhabdomyosarcoma [11–24]. It has been noticed that the number of studies examining the histology, behavior, and treatment results of middle ear tumors is low [25, 26]. The surgical procedure to be applied to the tumor is selected according to the size of the tumor, its location, and the general condition of the patient [27]. If the patient does not have persistent vertigo or extensive tumor growth, the watch and scan approach is one of the methods that can be preferred in treatment. En bloc resection is the preferred surgical method in middle ear cancers. Radiotherapy is additionally used in advanced lesions [28].

Conclusions

In this study, we aimed to present the case series of middle ear tumors along the period from 2010 till 2019. Tumors located in the middle ear, which generally cause similar clinical complaints, are rarely seen. It is necessary to evaluate the tissue histopathologically because it will affect the treatment and prognosis of the patients.

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Authors' contributions
SA has made substantial contributions to the conception or design of the work, and the acquisition, analysis, and interpretation of data. SA has approved the submitted version. SA has agreed both to be personally accountable for the author's own contributions and to ensure that questions related to the accuracy or integrity of any part of the work, even ones in which the author was not personally involved, are appropriately investigated, resolved, and the resolution documented in the literature. OS has made substantial contributions to the conception or design of the work, and the acquisition, analysis, and interpretation of data. OS has approved the submitted version. OS has agreed both to be personally accountable for the author's own contributions and to ensure that questions related to the accuracy or integrity of any part of the work, even ones in which the author was not personally involved, are appropriately investigated, resolved, and the resolution documented in the literature. OO has made substantial contributions to the conception or design of the work, and the acquisition, analysis, and interpretation of data. OO has approved the submitted version. OO has agreed both to be personally accountable for the author's own contributions and to ensure that questions related to the accuracy or integrity of any part of the work, even ones in which the author was not personally involved, are appropriately investigated, resolved, and the resolution documented in the literature.

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Availability of data and materials
The datasets generated and/or analyzed during the current study are not publicly available due to the reason why the data are not publıc but are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
The study was approved by Uludağ University Faculty of Medicine local Clinical Research Ethics Committee dated May 13, 2020 and numbered 2020-8/28. The committee's reference number is not available. Consent to participate is not applicable as it is a retrospective study.

Consent for publication
Verbal informed consent was obtained from the patients for publication of this article and accompanying images.

Competing interests
The authors declare that they have no competing interests.

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