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Case Report

Myxoma Arising from the Middle Ear: A Case Report

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The primary head and neck myxomas are rare, generally arising from the mandible, maxilla, and oral cavity. Other anatomical areas, such as cardiac myxomas, may also have metastases to the head and neck regions. The middle ear is an extremely rare location for myxomas. Myxoma slowly grows and is usually asymptomatic until it affects the surrounding structures. Surgical treatment is performed with a complete en bloc resection where possible. We report a case of a 42-year-old woman with myxoma arising from the right middle ear because of her tumor's rare anatomical region. Her main complaints were progressive fullness and loss of hearing which she felt for approximately 1 year on the right ear. High-resolution computed tomography (HRCT) revealed an isodense soft tissue mass localized in the right mastoid bone and the middle ear. The mass was totally removed by canal wall up tympanomastoidectomy. At the last follow-up examination on 36 months after the surgery, the patient was asymptomatic, and there were no signs of recurrence.

KEYWORDS: Myxoma, middle ear, head, neck, treatment

INTRODUCTION

Myxoma is a very rare benign tumor of the middle ear. The typical tumor development area is the atrium of the heart. Myxomas in the head and neck areas are mostly seen in the maxilla and mandibula. However, it may also develop in the auricular, ear canal, middle ear, and temporal bone. Other anatomical areas, such as cardiac myxomas, may also have metastases to the head and neck regions. The tumor grows slowly and is usually asymptomatic until it affects the surrounding structures in the ear [1-4]. Surgical treatment is performed with a complete en bloc resection where possible [2, 4].

In this case report, a 42-year-old woman with myxoma arising from the right middle ear was presented because of her tumor's rare anatomical region.

CASE PRESENTATION

A 42-year-old woman was admitted to our department with complaints of progressive fullness and loss of hearing which she felt for approximately 1 year on her right ear. On otoscopic examination, a polypoid mass in the external ear canal that was arising from the middle ear and covering the posterior inferior of the eardrum was revealed. Other examinations were unremarkable. High-resolution computed tomography (HRCT) revealed an isodense soft tissue mass filling the right mastoid and the middle ear. HRCT did not detect any bone destruction (Figure 1). The average air–bone gap (ABG) for four frequencies (0.5, 1, 2, and 4 kHz) on pure tone audiogram (PTA) was 26.25 dB on the right ear (Figure 2).

Written consent was obtained from the patient after informing about the procedure. Canal wall up (CWU) tympanomastoidectomy was performed by the retroauricular approach, and the pathological tissues in the middle ear and mastoid bone were removed. The origin of the mass was not clearly possible to determine. However, the mass was extending from the epitympanum to the mastoid cells via the aditus et antrum. Owing to the smooth surface of the polypoid mass and normal evaluation of the surrounding bone structures, macroscopically complete resection was performed, although not en bloc, as in conventional ear surgery. The ossic-

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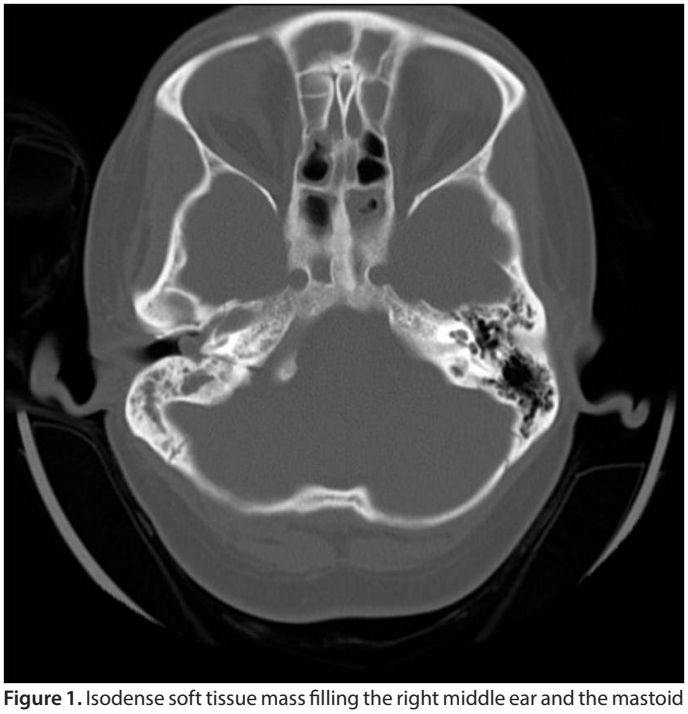

Figure 1. Isodense soft tissue mass filling the right middle ear and the mastoid in axial HRCT is being seen.
HRCT: high-resolution computed tomography

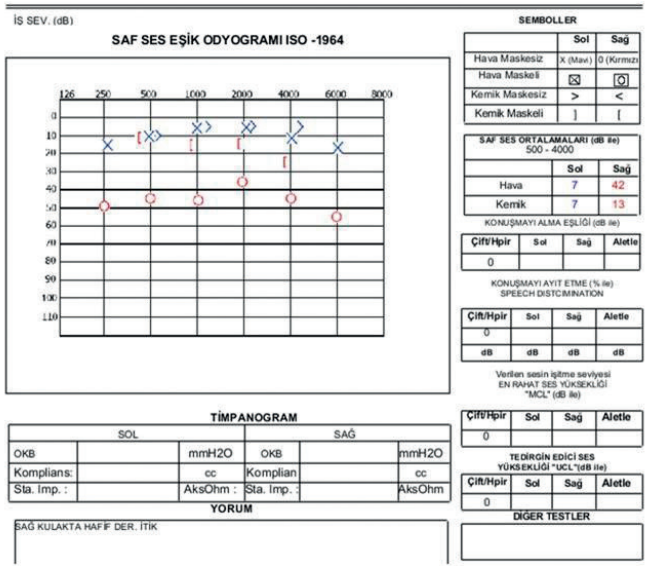

Figure 2. Preoperative pure tone audiogram of the patient.

ular chain integrity was preserved during dissection. The tympanic membrane areas in which the mass showed adhesion were excised together with the mass. Perforation of the tympanic membrane was reconstructed with perichondrium cartilage island graft.

Histopathologically, hematoxylin and eosin (H&E) staining revealed a keratinized multilayer squamous epithelium on the surface of the tissue and a clear myxoid area with regular boundaries under this area (Figure 3a, H&E ×40). Collagen fibers and spindle cells were exposed in the large myxoid stroma (Figure 3b, H&E ×200). Immunohistochemical S-100 staining was negative (Figure 3c, S-100 ×100). Positive staining was detected in several cells with CD34 (Figure 3d, CD34 ×100). Histopathological diagnosis was reported as myxoma.

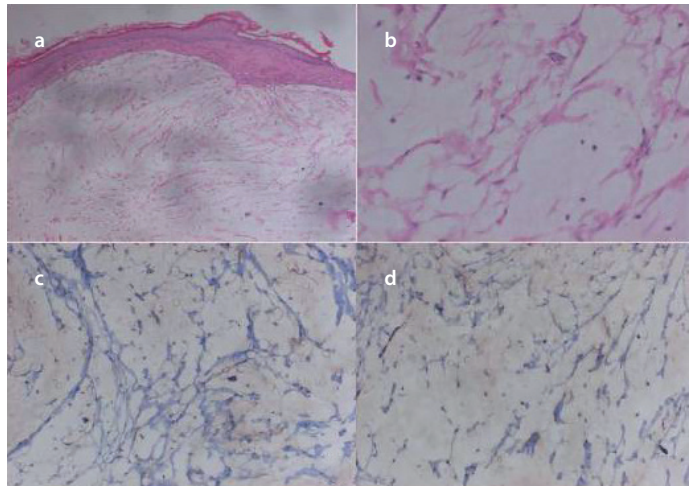

Figure 3. a-d. Hematoxylin and eosin (×40) (a). Hematoxylin and eosin staining collagen fibers and spindle cells are being seen in the large myxoid stroma ((×200) (b). Immunohistochemical negative staining with S-100 is being seen (×100) (c). Immunohistochemical positive staining in several cells with CD34 is being seen (×100) (d).
H&E: hematoxylin and eosin

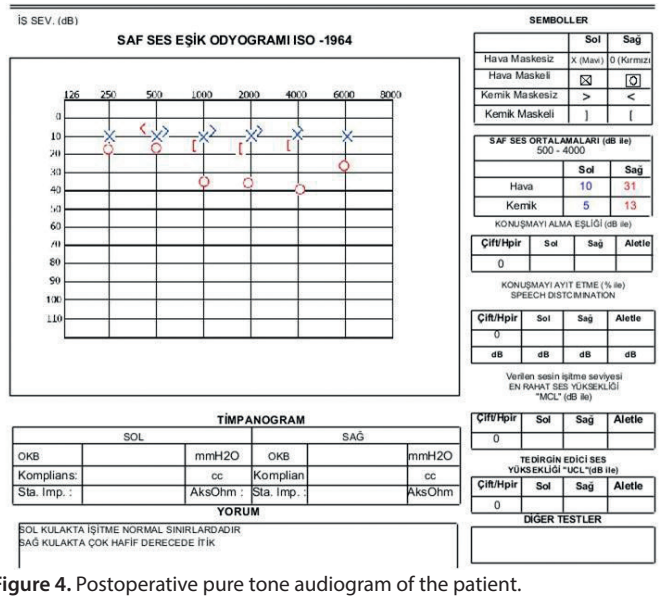

Figure 4. Postoperative pure tone audiogram of the patient.

At the last follow-up examination on 36 months postoperatively, the patient did not have any complaints and signs of recurrence. The average ABG for four frequencies (0.5, 1, 2, and 4 kHz) on PTA was 16 dB on the right ear (Figure 4).

DISCUSSION

Myxoma is a benign tumor originating from the mesenchymal tissue [1]. The typical tumor development area is the atrium of the heart [3]. However, it can also be arisen from the bone and other soft tissues [2]. Myxoma may be seen at any age, most frequently in the age of 20–30 years, with no gender dominance [4].

The primary myxomas of the head and neck areas are rare, generally seen in the mandible, maxilla, and oral cavity. Myxoma of the ear is so rare. Although myxoma is not classified as an ear tumor histologically by the World Health Organization, there are several reports of ear myxoma in the English literature [2]. The primitive embryonic

mesenchyme in the middle ear cavity is thought to be the origin of the tumor [2]. Sareen et al. [5] found 11 myxoma cases originating from the temporal bone in the literature and presented their own cases. Charabi described a case with temporal bone myxoma without intracranial extension [6].

Myxoma grows slowly in the ear and remains asymptomatic unless it stimulates the facial nerve or vestibular organ [3]. The main complaint in the presented case was fullness and hearing loss on the right ear.

Myxomas can also be a component of the Carney complex. Cutaneous, cardiac myxomas, schwannomas, and endocrine tumors are characteristics of this autosomal dominant disease. Cardiac myxomas are potentially fatal, so recognition of this disease is important [4, 7-9]. Briassoulis et al. [9] presented a 23-month-old patient with a diagnosis of Carney's complex. In their case, myxoma was reported in the ear lobule. In our case, there was no other abnormality on systemic examination.

High-resolution computed tomography and magnetic resonance imaging are important to determine the extension and specificity of the lesion, to help surgical planning, and to narrow the differential diagnosis [4, 7]. Radiological features of myxomas have been reported in several publications. The contrast enhancement and vascularity pattern of the tumor is variable; it can be seen as iso or hypodense mass [4]. In our case, isodense soft tissue mass filling the right middle ear and the mastoid in HRCT was determined.

It is important to distinguish the true myxoma, a benign lesion, from some soft tissue malignant tumors that show myxomatous degeneration. Nuclear atypia, mitotic figures, and also the incidence of high chromosomal abnormalities in the H&E staining sections, more pronounced vascularity and higher cellularity, are supportive in the recognition of these malignant tumors. Malignant myxoid tumors typically arise from the extremities and trunk [2].

The pathological diagnosis is confirmed by the presence of a true tumor consisting of satellite cells in the loose mucoid stroma. Myxomas have lobular pattern and separated to each other with connective tissue [1, 6]. Histologically, the tumor contains acid mucopolysaccharide-rich myxoid matrix [3].

More malignant lesions, such as myxoid liposarcoma, fibromyxoid sarcoma, myxoid chondrosarcoma, chondroid sarcoma, myxoid rhabdomyosarcoma, and meningioma, should be considered in the differential diagnosis [4].

Although it is a benign tumor, it is locally aggressive [4]. Large bone erosion was described by Nagatani et al. [1] in the pituitary fossa myxoma and Osterdock et al. [4] in the temporal bone myxoma.

Management of the myxomas in the bone is more difficult, because they are diagnosed late and require the resection of a bone segment [2]. If the radical resection is not appropriate due to the anatomical localization, the gelatinous structure of the myxoma makes the treatment challenging [4, 6]. Myxomas arising from the temporal bone with intracranial extension make radical resection impossible. Otherwise, the morbidity rate will be unacceptable. In these cases, fragment resection may be appropriate [4]. Radiotherapy and chemotherapy are not effective [2, 4, 5, 8].

The local destructive nature of these tumors complicates the accurate assessment of tumor boundaries. High recurrence rates of 25% have been reported in enucleation cases [8]. Radical excision is advocated in appropriated cases [4]. Soft tissue myxomas developing in the head and neck regions are easily removed with minimal morbidity [10].

In our presented case, the biopsy could be extracted from the mass before the operation. However, we did not need a biopsy because we did not suspect any malignancy or vascularity in both otoscopic and radiological evaluation of the lesion.

En block resection should be the treatment option. However, this may not be possible depending on the anatomical localization of the lesion. In this presented case, owing to the smooth surface of the polypoid mass and normal evaluation of the surrounding bone structures, a macroscopically complete resection was performed, although not en bloc, as in conventional ear surgery. Any bone destruction during surgery was not detected on HRCT.

Recurrence after surgical resection can be seen between 3 months and 10 years [4]. Long-term follow-up is required [7].

CONCLUSION

The middle ear is an extremely rare location for myxomas. Myxoma grows slowly and is usually asymptomatic until it affects the surrounding structures. The treatment is performed surgically with complete block resection. Long-term follow-up is required because of the high recurrence rate. In our presented case, the mass was removed by CWU tympanomastoidectomy. At 36 months postoperatively, the patient did not have any complaints or signs of recurrence.

Informed Consent: Written inform consent was obtained from the patient who participated in this study.

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Conflict of Interest: The authors have no conflict of interest to declare.

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