The Journal of International Advanced Otology

J Int Adv Otol 2020; 16(1): 134-7 • DOI: 10.5152/iao.2019.6709

Case Report

Primary Liposarcoma with Cholesteatoma in Mastoid

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Cite this article as: Sato MP, Saito K, Fujita T, Seo T, Doi K. Primary Liposarcoma with Cholesteatoma in Mastoid. J Int Adv Otol 2020; 16(1): 134-7.

Liposarcoma is a soft tissue neoplasm that commonly develops in the lower extremities and rarely in the head and neck region. Herein, we report the case of a patient with primary liposarcoma that was detected in the mastoid antrum during staged tympanoplasty for cholesteatoma. The tumor adjacent to the attic cholesteatoma was resected completely, and the pathological diagnosis was that of myxoid-type liposarcoma. Because positron emission tomography after the surgery showed no signs of tumor remnants or systemic metastasis, a second-stage surgery was performed 8 months after the first surgery. After confirming that there was no recurrence, tympanoplasty type III with interposition between the stapes and malleus and canal reconstruction was performed. No recurrence was observed for 5 years, and to date, good hearing has been maintained. This is the first report on long-term follow-up of a patient with liposarcoma in the mastoid antrum.

KEYWORDS: Liposarcoma, mastoid, cholesteatoma, soft tissue neoplasm

INTRODUCTION

Liposarcoma arising from the fascia, muscle, or adjacent mesenchymal tissue is commonly identified in the retroperitoneum and lower extremities, but it is extremely rare in the head and neck region [1]. Three cases of primary liposarcoma in the mastoid have been reported [2-4]. Herein, we present a case of myxoid-type liposarcoma in the mastoid antrum incidentally found while performing first-stage tympanoplasty procedure for cholesteatoma with a long-term follow-up of >5 years.

CASE PRESENTATION

A 52-year-old male was referred to our hospital with the chief complaint of hearing loss and ear fullness in the right ear. Otoscopy revealed dry attic retraction without squamous debris. Pure tone audiometry showed a conductive hearing loss of 55 decibels (dB) in the right ear. Computed tomography (CT) images revealed a soft tissue shadow in the middle ear that extended to the mastoid with erosion of the middle cranial fossa (Figures 1a, b). Images obtained via T2-weighted magnetic resonance imaging (MRI) showed a slightly hyperintense signal and those obtained via diffusion-weighted imaging showed a moderately intense lesion in the same area (Figures 1c, d). Therefore, we suspected cholesteatoma with cholesterol granulation on the right side.

The first-stage tympanoplasty was performed in 2013. Cholesteatoma eroded the scutum and invaded the head of the malleus, extending to the malleoincudal joint. The body of the incus was fixed because of the granulation. After removing the incus, we proceeded to perform mastoidectomy. Observation from the mastoid side revealed that a smooth, soft, and yellowish tumor was trapped from the attic to the antrum (Figure 2a). We were able to resect the tumor completely because it had not adhered to the neighboring tissues (Figure 2b). Although the middle fossa plate had thinned, the dura was not exposed. This thinning was presumed to have been caused by this tumor. The attic cholesteatoma was also removed completely, and silastic sheets (Kaneka Medix Corporation, Osaka, Japan) were placed in the middle ear for ventilation. Canal-wall-down tympanoplasty with canal reconstruction was finally performed. The pathological diagnosis was that of attic cholesteatoma and myxoid liposarcoma in the antrum. In the latter, plexiform capillary vascular networks were observed, and lipoblasts were present in the specimen (Figures 3a, b).

Postoperative positron emission tomography (PET) images showed no abnormal hot spots. Because the patient opted for observation without postoperative chemoradiotherapy, we performed the second stage of the surgery as planned to check for recurrence and improved hearing 8 months after the initial surgery. At surgery, some granulations were observed at the same site. Rapid

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Submitted: 25.02.2019 • **Revision Received:** 03.04.2019 • **Accepted:** 09.04.2019 • **Available Online Date:** 27.06.2019

Available online at www.advancedotology.org

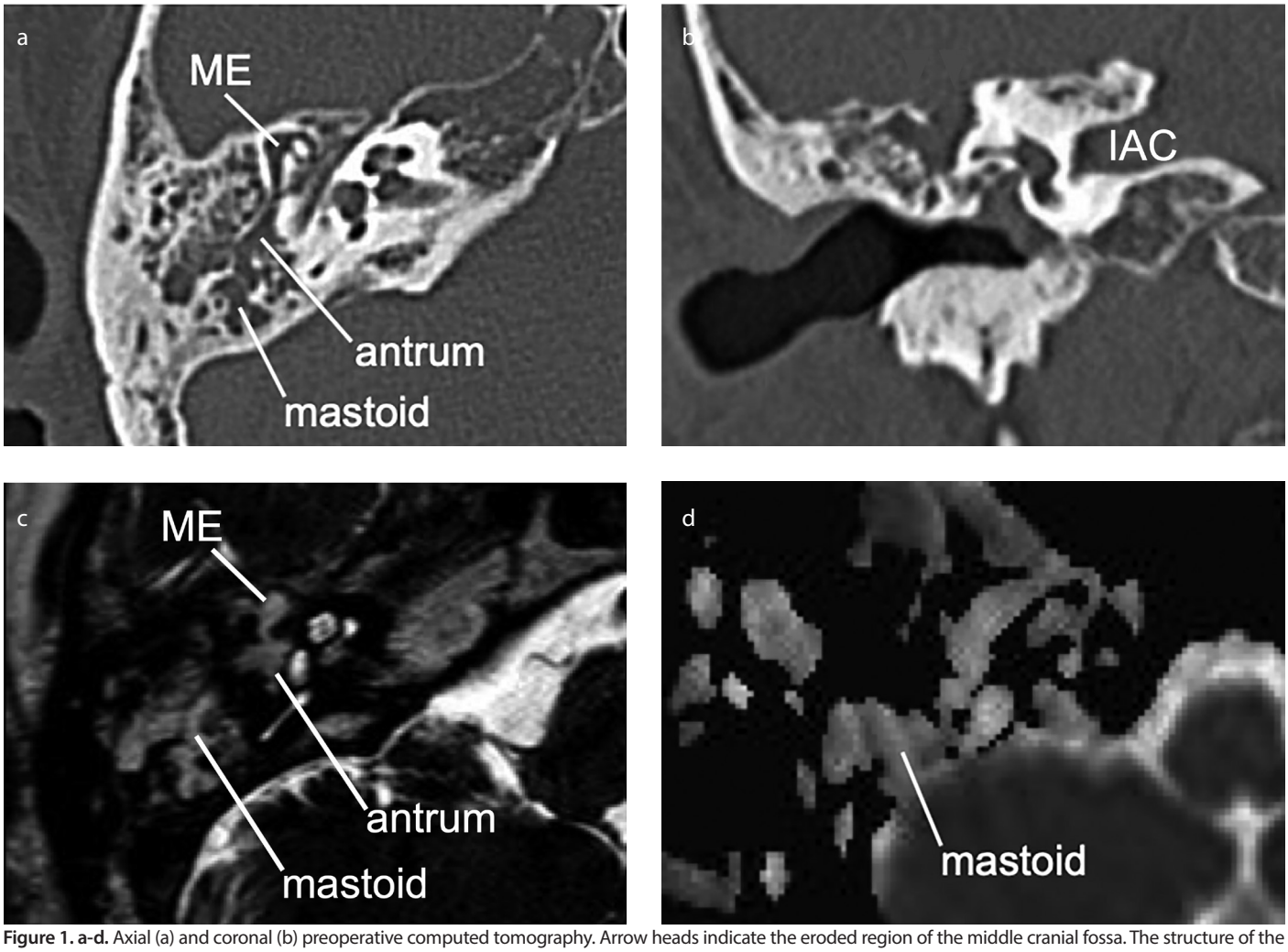

Figure 1. a-d. Axial (a) and coronal (b) preoperative computed tomography. Arrow heads indicate the eroded region of the middle cranial fossa. The structure of the mastoid seemed to remain almost normal. Coronal T2-weighted (c) and diffusion-weighted (d) magnetic resonance imaging shows slight hyperintense signal in the middle ear, antrum, and mastoid. ME: middle ear, IAC: internal auditory canal.

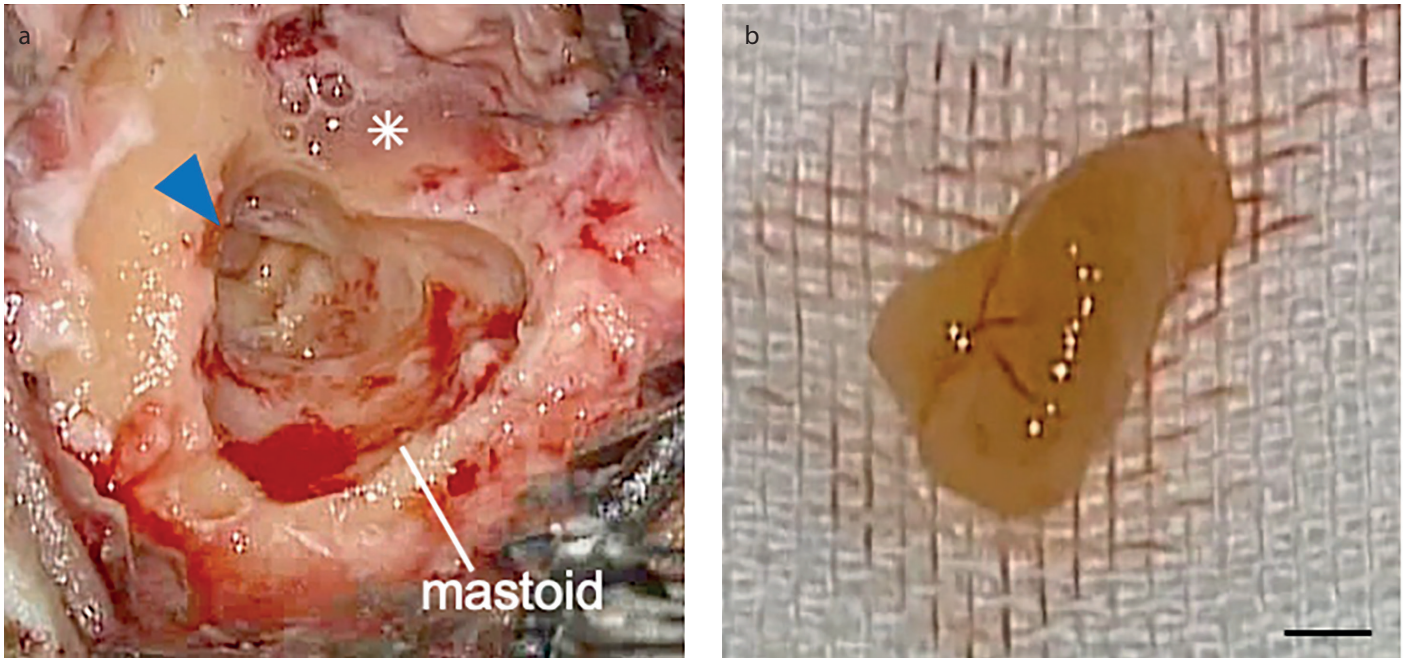

Figure 2. a, b. The intraoperative picture after mastoidectomy (a). Arrow head indicates the tumor lying in the mastoid antrum. Asterisk shows the external auditory canal. The excised tumor is shown in (b). The black bar indicates 2.5 mm.

Table 1. Summary of other reports of mastoid liposarcoma

Case	First Author	Age (years)	Chief complaint	Type	Treatment	Recurrence
1	P.N. Agarwal 1975 [2]	4 Facial palsy	Otorrhea	Round cell	Surgery	Unknown
2	A.P. Coatesworth 1996 [3]	85 Facial palsy	Otorrhea	Round cell	None	Unknown
3	Seo T. 2007 [4]	69	Otorrhea Otalgia	Well-differentiated	Surgery	None

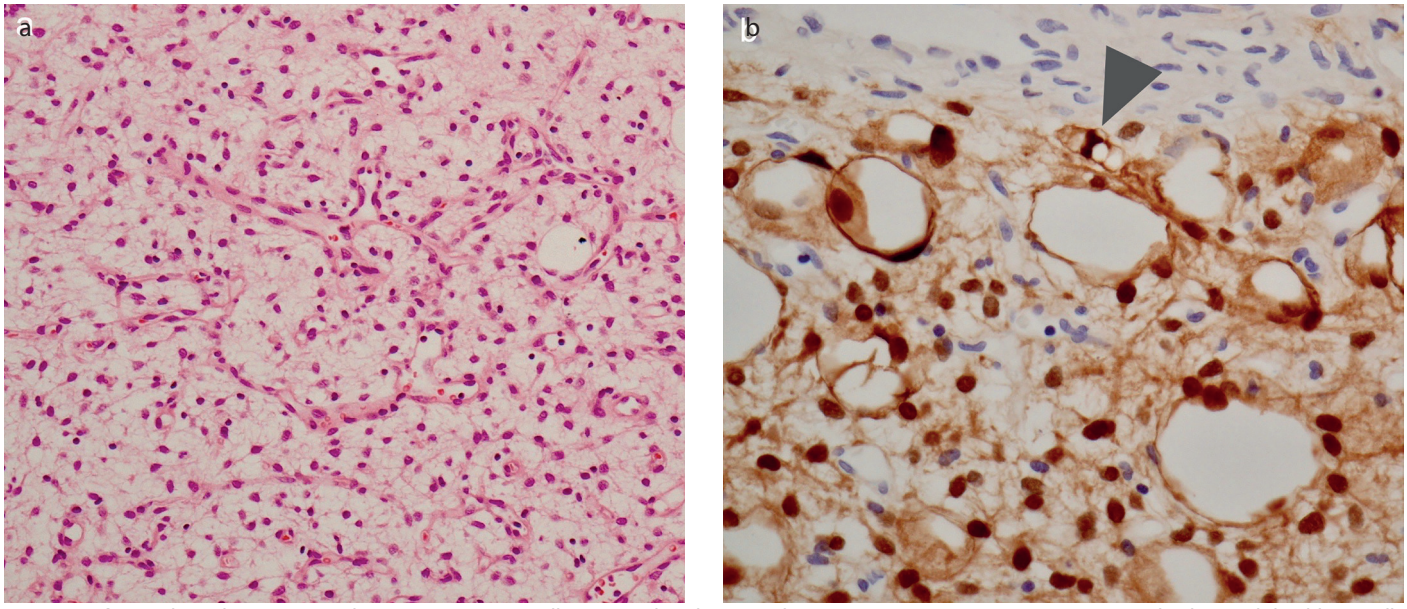

Figure 3. a, b. Histological examination shows an extensive capillary network on hematoxylin-eosin staining (a, × 200). S-100 staining clearly reveals lipoblastic cells (arrow head) (b, × 400).

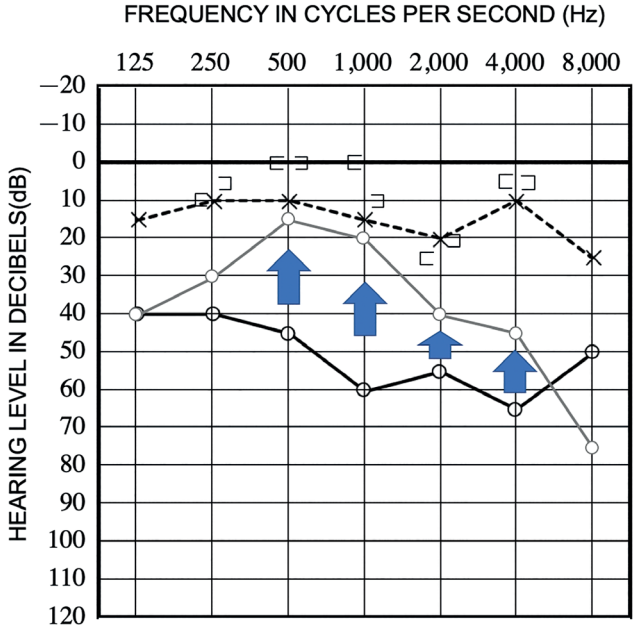

Figure 4. Pure tone audiogram. Hearing in the right ear at the first visit (darker line) improved after the second surgery and remained well for 5 years.

pathological diagnosis during surgery revealed no malignancy, and the final pathological results revealed no tumor recurrence. Furthermore, hearing improved owing to tympanoplasty type III with interposition between the stapes and malleus and canal reconstruction (Figure 4). There has been no evidence of recurrence on images obtained via CT, MRI, or PET in the 5 years after the initial surgery, and good hearing has been maintained to date.

DISCUSSION

This is the fourth reported case of liposarcoma in the mastoid antrum. We have reviewed the English literature on primary liposarcoma in the mastoid antrum (Table 1). In one case out of the three previously reported cases, early detection and treatment were possible owing to a complication of cholesteatoma, which was similar to our case [2]. This patient underwent surgery twice, as did the patient in our study, and was followed up for 2 years after the first surgery and no recurrence was detected. Early detection of tumor remnants and recurrence in the mastoid is very challenging, and a second surgery would be necessary for better management. The surgical technique used in canal reconstruction depends on the situation. In our case, canal reconstruction was performed because the rapid pathological diagnosis at the second surgery showed no recurrence. When recurrence is suspected, canal-wall-down tympanoplasty would be better in terms of long-term follow-up. Local recurrence commonly occurs within the first 6 months after initial surgery but may sometimes be delayed for 5–10 years [5]. Long-term observation is necessary with the use of imaging modalities, such as CT, MRI, and PET.

The preoperative diagnosis for such a small lesion is very difficult. Although we checked the preoperative CT and MRI scans retrospectively,

liposarcoma could not be determined clearly. This situation leads to two problems in treatment. The first is the safety margin of the mastoid lesion. Mouret et al. [6] reported that the safety margin requires a distance of at least 2 cm around the lesion because sometimes, satellite nodules are present around it. The second is a precise diagnosis by postoperative pathology. This case was assumed to be that of cholesteatoma with suspicion of cholesterol granuloma at first. Because middle ear tumors sometimes mimic or are accompanied by cholesteatoma [7], pathological examination for cholesteatomas as well should be routinely performed with great care. The pathological examination has been performed for cholesteatoma at our facility, so that a precise diagnosis is possible, as in such a rare case.

The prognosis for liposarcoma depends on the pathological type, which can be divided into five categories in accordance with Enzinger's classification: (i) well-differentiated type, (ii) myxoid type, (iii) round cell type, (iv) dedifferentiated type, and (v) pleomorphic type; (i) and (ii) are regarded as low-grade tumors, whereas the others are considered as high grade. In our case, appropriate local control by performing two surgeries appeared effective because low-grade tumors do not seem to involve metastases.

There can be two options for the treatment of recurrent mastoid liposarcoma. The first is temporal bone resection, and the second is radiotherapy. We planned the latter if recurrence was detected during the second surgery. Postoperative radiotherapy is useful because the sensitivity is better, especially for myxoid-type tumors, as in our case [8]. Radiation-induced sarcoma had a prevalence ranging from 0.02% to 0.55% [9], and hearing could be lost with mastoid lesions. Case 2 in Table 1 was actually induced by radiation. Patel et al. [10] reported that chemotherapy was effective in 44% of 21 cases of myxoid-type liposarcoma.

CONCLUSION

We reported a rare case of myxoid liposarcoma in the mastoid antrum with cholesteatoma. The two-stage surgery prevented recurrence and maintained good hearing for >5 years. Careful pathological examination for cholesteatoma is essential for a precise diagnosis.

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

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – M.P.S., K.D.; Design – M.P.S., T.F., K.D.; Supervision – T.F., T.S., K.D.; Resource – M.P.S., K.S.; Materials – M.P.S., K.S.; Data Collection and/or Processing – M.P.S., K.S., T.F.; Analysis and/or Interpretation – M.P.S., T.F., T.S.; Literature Search – M.P.S., T.S.; Writing – M.P.S., K.S., T.F.; Critical Reviews – M.P.S., K.S., T.F., T.S., K.D.

Acknowledgements: We are grateful to Prof. Takao Sato for his cooperation for pathological diagnosis.

Conflict of Interest: The authors declare that they have no conflicts of interest.

Financial Disclosure: The authors declared that this study has received no financial support.

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