Chondroblastoma of the temporal bone

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Running Head: temporal bone chondroblastoma

Abstract

CONCLUSIONS: Temporal bone chondroblastomas are extremely rare and aggressive, but the outcome after appropriate surgical treatment is favorable. From the review, it may be particularly important to deal with tumors that involve the temporomandibular joint (TMJ), which could affect the long term outcomes, as well as tumor recurrence.

OBJECTIVE: To report a case of chondroblastoma and to review their presentation.

BACKGROUND: Chondroblastomas are highly destructive tumors that are derived from immature cartilage cells. The occurrence of this tumor in the temporal bone or skull base is uncommon. Approximately 70 cases have previously been reported, several of which have involved the TMJ.

CASE DESCRIPTION: We currently report the case of a 67-year-old woman who presented with right-sided mixed hearing loss, a right external auditory canal mass, ear fullness, otalgia, bloodstained otorrhea, and pain around TMJ, associated with difficulty in opening the mouth. CT and MRI revealed a mass involving the TMJ, infratemporal fossa, and pterygopalatine fossa. The patient underwent tumor resection via an infratemporal fossa approach type B. Gross total tumor removal was achieved, with no facial nerve paralysis or other complications observed after surgery. No recurrence or residual tumors were observed on CT and MRI, even after seven and a half years of follow-up.

Introduction

Chondroblastomas are rare benign bone tumors that represent approximately 1% of all primary bone tumors. Most of these tumors commonly arise in the epiphysis of the long bones, and tumors of the temporal bones are extremely rare [1]. A chondroblastoma of the temporal bone was first reported by Denko and Krauel in 1955, and since then, around 70 cases have been reported [2-4]. We report a case of chondroblastoma of the temporal bone, and review the literature to discuss the clinical presentation, radiological and pathological findings, as well as surgical management strategies.

Case report

In February 2003, a 67-year-old woman presented to our institute with a 5 month history of Right sided hearing loss. She also complained of tinnitus, otalgia, bloodstained otorrhea and temporomandibular joint (TMJ) pain on movement. However, she had no dizziness, facial palsy or other cranial nerve involvement. Upon otoscopic examination, there was a mass noted in the external auditory canal. The mass almost totally occluded the lumen of the external auditory canal, and the lesion allowed for only a limited view of the postero inferior part of the tympanic membrane (Figure 1). An audiogram showed a 56.7 dB mixed hearing loss with a 31.7 dB air-bone gap in the right ear.

A CT scan showed a solid, dense mass with diffuse calcification. The expansive growth had invaded the external auditory canal and extended into the middle ear with bony erosion (Fig. 2A). It also extended medial to the TMJ, anteriorly to the pterygopalatine fossa, and superiorly to erode the bony floor of the middle cranial fossa

(Fig. 2B, 2C). The tumor had a non-homogenous appearance and low intensity on T1 weighted MRI and showed iso to high intensity on T2 weighted MRI. The tumor had invaded TMJ capsule and middle cranial fossa superiorly (Fig. 3). The internal carotid artery was not involved.

The patient underwent resection via an infratemporal approach type B in March 2003. A cranio-temporal incision was performed and the external auditory canal was transected and closed. The extra temporal facial nerve was exposed in the parotid and a subtotal petrosectomy was performed. The tumor had involved the antero medial part of the bony external auditory canal and occupied the mesotypanum and anterior attic. Additionally, the tumor had invaded the middle cranial fossa dura and involved the glenoid fossa but spared the condyle. The skin of the bony external auditory canal, the tympanic membrane, the malleus, and the incus were removed after disarticulation of the incudostapedial joint. The facial nerve was skeletonized and the inner ear was preserved, while the vertical portion of the internal carotid artery was identified and skeletonized. The capsule of the TMJ was detached and removed but since the condyle was not involved, it was left in place. The middle meningeal artery and the mandibular nerve were coagulated and cut, and the bony eustachian tube was drilled out, identifying the horizontal part of the carotid artery. Although the tumor was adherent to the middle cranial fossa dura, it was easily detached and was removed completely. Lastly the eustachian tube was closed and the operative defect obliterated with abdominal fat.

The patient had an uneventful postoperative period without any signs of facial palsy after surgery or any other complications, and was discharged 3 days later. The resected tumor showed histological findings consistent with chondroblastoma. Her 1

year follow up CT revealed that the surgical defect was obliterated with adipose tissue as evidenced by her latest MRI, and she is still disease free seven and a half year later (Figure 4).

Discussion

Clinical findings

Chondroblastomas typically occur in the long bone epiphyses of children, and are uncommon in patients over 25 years of age [5, 6]. Chondroblastoms involving the temporal bones are uncommon, and only 70 cases have been reported, with only 45 having detailed pathologic and clinical treatment data. The common location of a temporal bone chondroblastoma is at the squamous portion, due to its cartilaginous origin [7, 8]. The tumor is also anatomically attached to the TMJ region and presumably originates from the TMJ articular hyaline cartilage [9]. The characteristic growth pattern of the tumor may result from embryonal cartilaginous rests entrapped in the tympanosquamous suture line in the temporal bone. The suture is anterosuperior to the external auditory canal and anatomically very close to TMJ [10]. Most patients with temporal bone chondroblastomas were older than those with tumors at the usual site, the ages ranged from 8 years to 85 years old, with a mean of 39.4 [4, 11]. Of the 45 patients, 20 were male and 25 were female, and there was a variety of symptoms. 34 patients (76%) presented with otologic complaints, including hearing loss, tinnitus, otalgia, otorrhea and vertigo, and 12 patients (27%) complained of a temporal mass or swelling, whereas 11 patients (24%) presented with difficulties of mastication and TMJ symptoms [3, 4, 8, 12]. In terms of neurological complaints, 6 patients (13%) presented with headache or seizures, 7 patients (16%) presented with facial palsy, and 1

patient presented with a lower cranial nerve dysfunction [4]. The duration of symptoms was known in 41 cases, and ranged from 20 days to 10 years, with a mean of 15 months [13]. Our patient was symptomatic for 5 months, and was older than the mean age that has been reported. The patient exhibited the typical symptoms of chondroblastoma including hearing loss, tinnitus, otalgia, otorrhea and pain on TMJ movement. Compared to the mean length of follow-up (3.6 years) from 36 cases, our patient was followed up for a relatively long period of seven and a half years after surgery [4, 14].

Radiological findings

Radiologically, temporal bone chondroblastomas are solid or cystic masses that are characterized as osteolytic lesions, sometimes with spotty calcification. Calcification within the lesion has been documented in 20 to 50% of cases and can be identified well with CT [4, 6, 11, 15, 16]. However, although diffuse calcification patterns are uncommon, our case displayed calcification in almost the entire tumor unlike the CT findings of previously reported cases of temporal bone chondroblastoma. The appearance of chondroblastomas is variable in MRI and various patterns of enhancement, either peripheral, homogenous or heterogenous, have been seen with the use of gadolinium [4, 17]. There is usually low intensity on T1 images, and low to high intensity on T2 images and our case displayed similar findings consistent with previous reports.

Pathological features

On the basis of clinical and radiographic examination results, malignant lesions

such as chondrosarcoma, osteosarcoma, malignant fibrohistocytoma as well as benign osseus tumors such as giant cell tumor, osteoma and fibrous dysphasia must be considered. A chondroblastoma is composed primarily of immature chondroblasts with focal ‘‘chicken-wire’’ calcification, multinucleated giant cells, polygonal cells and acidophilic cytoplasm with the chondroid matrix [7, 8]. Immunoreactivity to S-100 has been widely used to assist in the differentiation of chondroblastomas from other pathological identities as S-100 positivity is present in 90% of chondroblastomas, and only 13% of giant cell tumors [18].

Surgical Treatment

Complete surgical resection is the standard procedure for the treatment of a temporal bone chondroblastoma. It has been reported that approximately 50% of patients with temporal bone chondroblastomas who were treated with curettage developed recurrent disease, requiring a secondary procedure, while the overall recurrence rate after total tumor removal is about 20%, according to the published literature [6, 7, 19, 20]. Bertoni et al. reviewed the published literature and found that TMJ extension was seen in 2 out of 21 patients (10%) [20]. In the published literature, we found that 16 out of the 45 patients (36%) had tumor invasion into the TMJ, and 5 out of 9 patients (56%) had TMJ involvement of the tumor in recurrent or residual cases (Table 1) [3-5, 7, 10, 12-17, 19]. The involvement of the TMJ and petrous bone is common in recurrent or residual cases. Several cases of temporal bone chondroblastomas need additional treatment or radiation therapy (because of recurrence or residual tumor). [4, 6, 14]

Because of the close anatomical relationship of a chondroblatoma to vital

neurovascular structures, aggressive attempts to completely remove the tumor without adequate exposure would be hazardous. Since chondroblastomas usually arise from the anterior temporal bone near the TMJ, adequate exposure of the TMJ is essential. A high recurrence rate of temporal bone chondroblastoma has been previously reported when using the via middle cranial fossa approach [3, 6]. Kurokawa et al. reported that surgical treatment of the tumors via a transzygomatic approach with mandibular condyle removal and exposure through the middle fossa approach is not adequate for removing temporal bone chondroblastomas. On the other hand, an infratemporal fossa approach provides adequate exposure for large tumors involving the TMJ. Infratemporal fossa approach type B is extralabyrinthine approach which is designed mainly for extra dural lesions involving the petrous apex and clivus including the horizontal segment of the internal carotid artery (Fig. 5). The posterior infratemporal fossa is accessible but the pterygoid process remains intact. Inftratemporal fossa approach type C is an anterior extension of the infratemporal fossa approach type B, in which the pterygoid process is drilled, and is used for exposure of the infratemporal fossa, pterygopalatine fossa, parasellar region, and nasopharynx. Both infratemporal fossa approaches provide good control of the TMJ. Some recent publications reported that excellent outcomes for temporal bone chondroblastomas can be achieved by preserving the TMJ or mandibular condyle through the infratemporal fossa approach [4, 13]. The key to the successful treatment of a temporal bone chondroblastoma might be the surgical management of the tumor obscured by the glenoid fossa and TMJ which can be accessed via an infratemporal fossa approach as our report.

Conclusion

We present a case of a patient with a chondroblastoma of the temporal bone and review the previously-published literature. A chondroblastoma of the temporal bone is an uncommon tumor, but the outcome after appropriate surgical treatment is quite favorable, and it is our opinion that proper access is necessary to deal with tumors involving the temporomandibular joint, which could be related to the long term outcome of the patient, as well as tumor recurrence.

Acknowledgements

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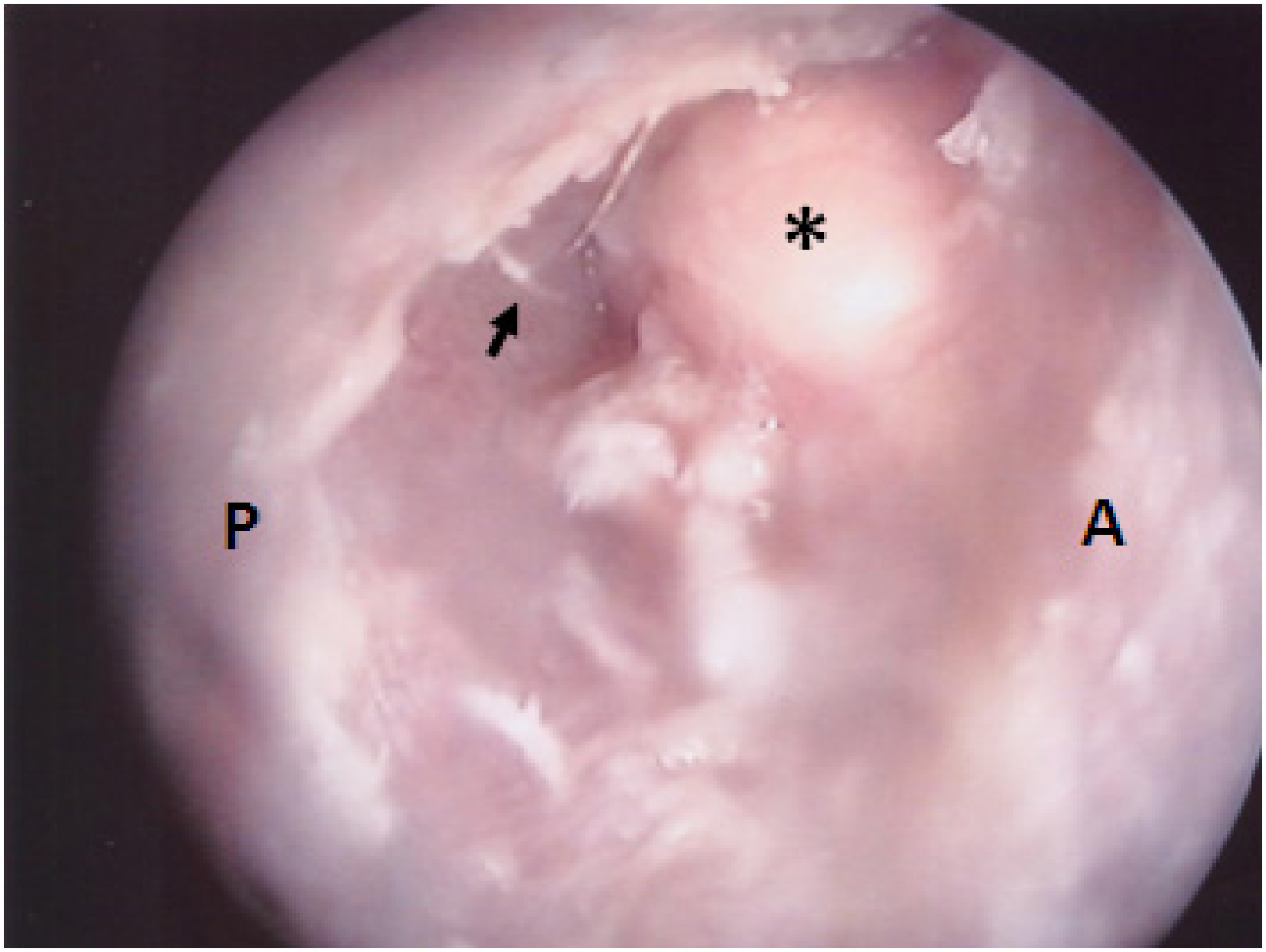
Table 1　Summary of temporal bone chondroblastoma cases: tumor invasion into temporomandibular joint (from the literature).

	Sex	Age	Symptoms	Site of Tumor	Treatment
Anim and Bakara, 1986	m	55	HL, Painful Swelling, Otorrhea	TMJ, Posterior End of Zygoma	Trans Mastoid
Tanohata et al., 1986	f	55	HL, Tinnitus, Otalgia, Headache	Anterior to TMJ, Squamous, MF	Resection
Horn et al., 1990	m	34	HL, Painful Swelling	Replaced Glenoid Fossa	Trans Mastoid and MFA
Politi et al., 1991	m	53	Swelling	Glenoid Cavity, Squamous	Curettage
Varvares et al., 1992	m	33	HL, Otalgia, Headache, TMJ Dysfunction	EAC, Petrous, TMJ	Trans Mastoid and Cervical
Leong et al., 1994	m	23	Ear Fullness on Mastication	Anterior to TMJ, MF	Trans Mastoid
Selesnick and Lenine, 1999	f	30	Otalgia, TMJ syndrome, Trismus	Lateral and Medial to Condyle, MF	Zygomatic, MFA
Watanabe et al., 1999	f	43	HL, Ear Fullness	Superior and Anterior to TMJ, Posterior Zygomatic Root, Squamous, MF	Trans Mastoid
Ishikawa et al., 2002	m	24	Arthralgia of TMJ, Painful Swelling, Difficulty of Mouth Opening	Pterygoid Fossa, TMJ, MF Dura	Zygomatic
Pontius et al., 2003	m	38	Otalgia, Otorrhea	EAC, TMJ, Petrous, Squamous	MFA, Trans Mastoid
Kurokawa et al., 2005	f	32	HL, Tinnitus, Limitation of Mouth Opening	Mandibular Fossa, Zygomatic Root, Middle Ear	Zygomatic
	m	27	HL, Tinnitus, Pain of TMJ	Mandibular Fossa , Zygomatic Arch	Zygomatic
Cabrera et al., 2006	f	38	Parotid Mass	TMJ, Parotid Gland	Excision
Kutz et al., 2007	f	62	HL, Otalgia, Ear Fullness	Condyle, Middle Ear, MF	ITFA
	f	39	Otalgia, Headache, Facial Palsy	Anterior to Condyle	ITFA
Moon et al., 2008	f	48	HL, Facial Palsy, Trismus	Sphenoid bone, Carotid Canal, Condyle, Inner Ear	ITFA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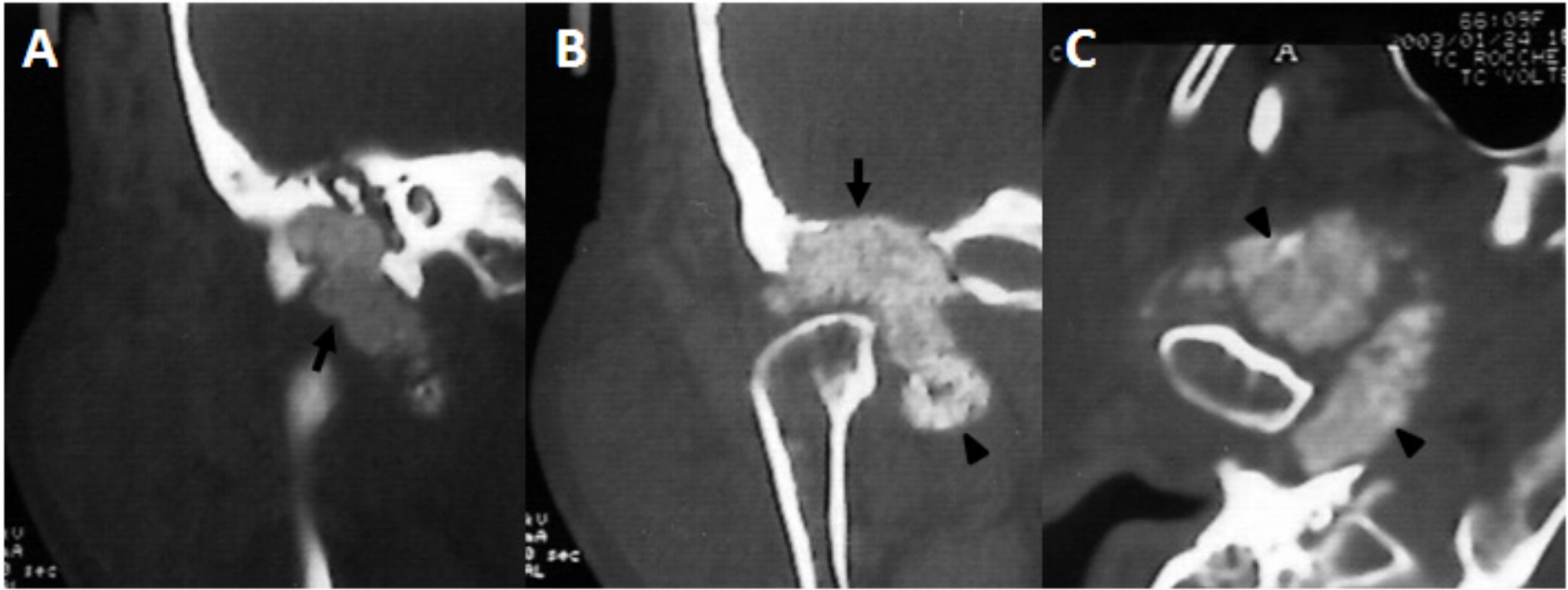
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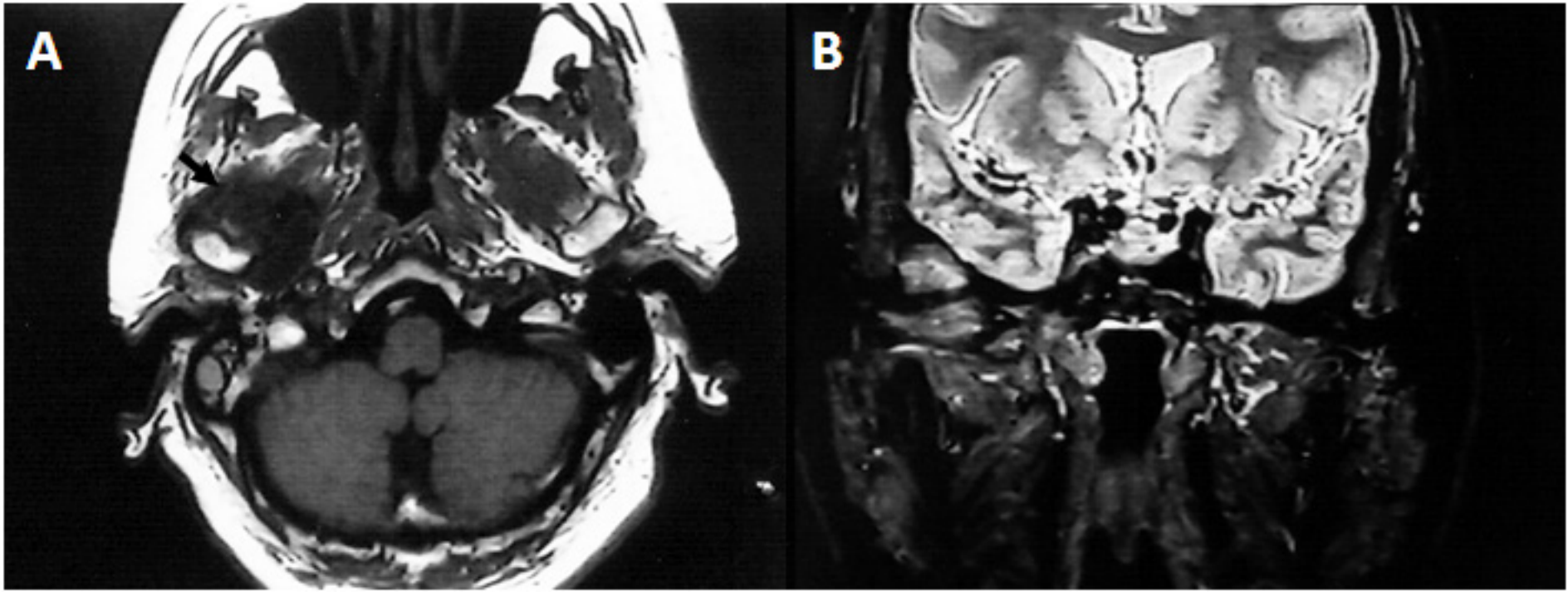
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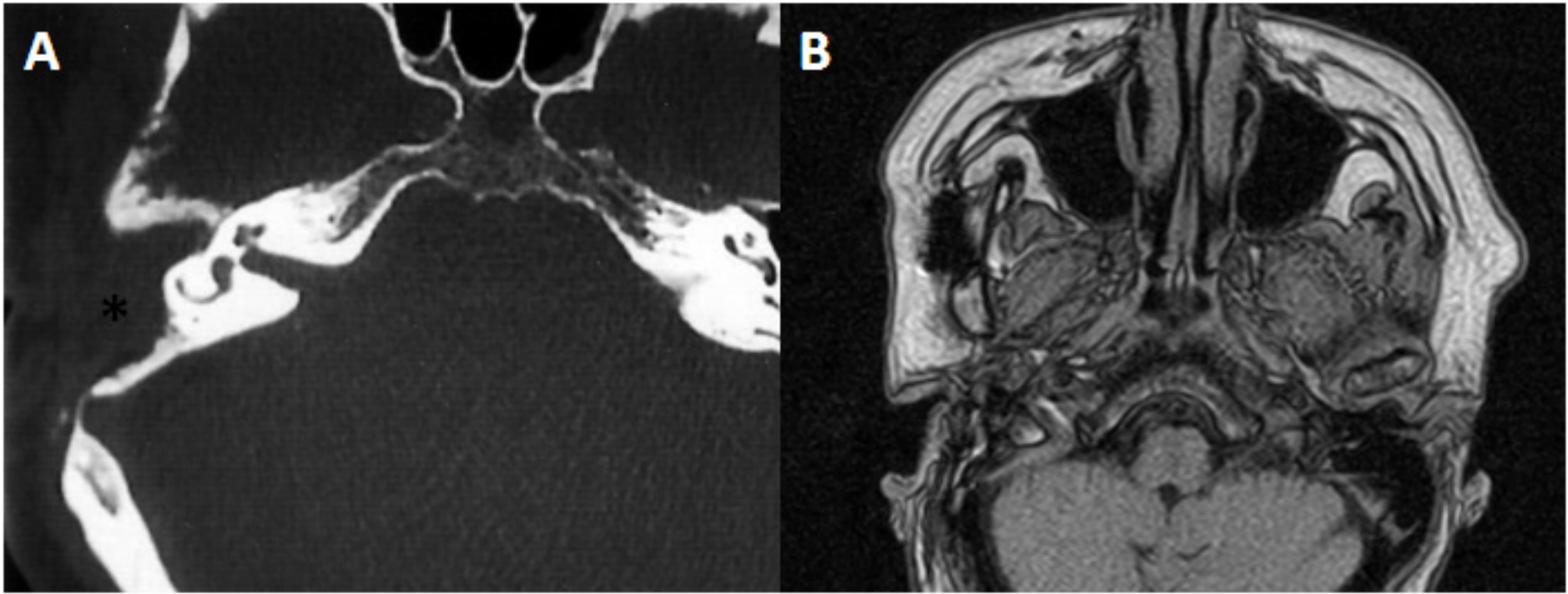
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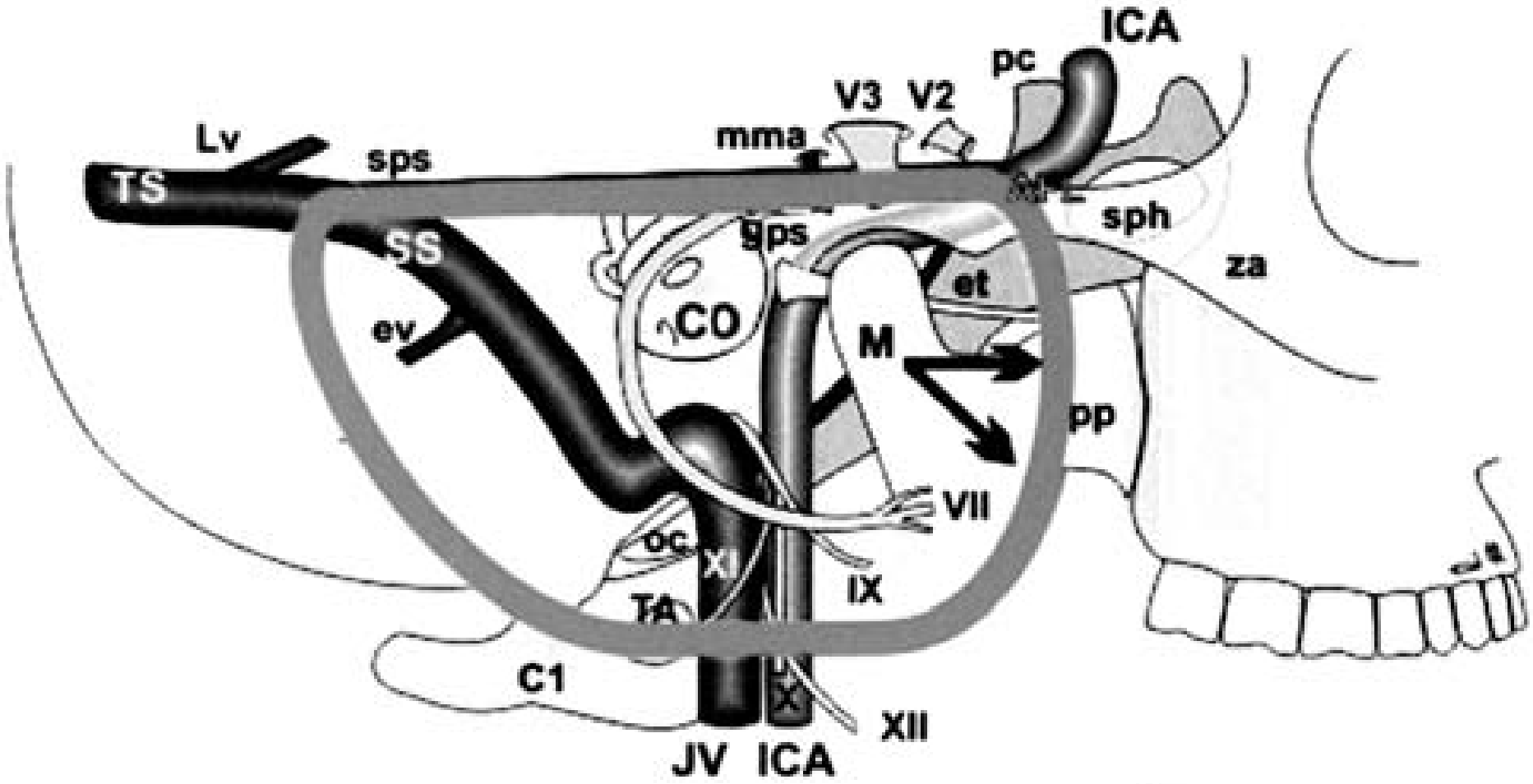
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