Case 14843
Chondromyxoid fibroma of the mastoid

Nicolas De Vos[1, 2]
David Creytens[3]
Maritte De Cock[4]
Filip Vanhoenacker[1, 2, 5]

1. Department of Radiology, Ghent University Hospital, Ghent University, Ghent
2. Department of Radiology, AZ Sint-Maarten Duffel-Mechelen
3. Department of Pathology, Ghent University Hospital, Ghent University, Ghent
4. Department of Otorhinolaryngology, AZ Sint-Maarten, Duffel-Mechelen
5. Department of Radiology, Antwerp University Hospital, Antwerp University, Antwerp University Hospital Antwerp

Section: Neuroradiology
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Patient: 21 year(s), female

Clinical History

A 21-year-old female presented at the emergency department with drooping of the left eyelid and left mouth corner. Neurological examination showed a left lower motor neuron facial nerve palsy. Otoscopy showed narrowing of the left external auditory canal.

Imaging Findings

Magnetic resonance imaging (MRI) demonstrated a space-occupying lesion in the mastoid portion of the left temporal bone. On T1-weighted imaging (T1-WI), the lesion was of low signal intensity (SI) (Fig. 1A). On T2-weighted imaging (T2-WI), the lesion displayed heterogeneous intermediate to high SI (Fig. 1B). Diffusion-weighted images and apparent diffusion coefficient maps showed no significant diffusion restriction (Fig. 2). After intravenous administration of gadolinium-based contrast medium, the lesion enhanced heterogeneously, with marked enhancement in the areas which were of intermediate SI on T2-WI, and absence of enhancement in the areas which were of

high SI on T2-WI (Fig. 3)
Cone-beam computed tomography confirmed the presence of a destructive osteolytic space-occupying lesion in the left mastoid (Fig. 4). The lesion eroded the facial canal, explaining the facial nerve palsy of the patient. The lesion also extended into the external auditory canal, explaining narrowing of this canal on otoscopy.

Discussion

CMF is an uncommon benign bone tumor of cartilaginous origin (1, 2, 3). It is usually found in the metaphyses of the long bones of the lower extremities, especially the proximal metaphysis of the tibia (4). CMF is also seen in the flat bones and bones of hands and feet. There is a slight predilection in males and a peak incidence in the second and third decades. CMF is rarely encountered in the skull base and is extremely rare in the mastoid portion of the temporal bone (5, 6). This case is only the eleventh identified in the literature (Table 1).
Clinical perspective: Patients with CMF of the appendicular skeleton primarily complain of pain and soft tissue swelling. Symptoms of CMF of the mastoid include hearing loss (7-11), facial nerve palsy (4, 5, 12), otalgia (2, 4, 9), and vertigo (6, 8).
On plain radiographs, CMF of the appendicular skeleton is often seen as an eccentric radiolucent lesion with a well-defined sclerotic margin (13). On MRI, CMF of the appendicular skeleton shows low SI on T1-WI and high SI on T2-WI. After intravenous administration of gadolinium-based contrast medium, peripheral nodular enhancement is seen in 70 % of lesions, while diffuse contrast enhancement is seen in 30% (14). For CMF of the mastoid, (cone-beam) CT and MRI are helpful in elucidating bone and soft tissue extension, respectively. On (cone-beam) CT, CMF of the mastoid is seen as a destructive, osteolytic, space-occupying lesion. Intratumoral calcifications are more frequently seen in CMF of the mastoid compared to CMF of the appendicular skeleton. On MRI, CMF of the mastoid shows low SI on T1-WI and heterogeneous intermediate to high SI on T2-WI. This heterogeneity can be attributed to the varying composition of chondroid, myxoid, and fibrous elements. Sometimes, cystic or hemorrhagic foci are seen. After intravenous administration of gadolinium-based contrast medium, there is heterogeneous enhancement, with clear enhancement in the areas which show intermediate SI on T2-WI, and absence of enhancement in the areas which show high SI on T2-WI. The latter are probably composed of predominantly chondroid tissue. Compared to CMF of the appendicular skeleton, CMF of the mastoid can occur at an older age, contains more intratumoral calcifications and shows more heterogeneous SI on T2-WI. Since both radiological and histopathological findings often show considerable overlap with other diseases, including endolymphatic sac tumors and malignant cartilaginous tumors, the diagnosis of CMF remains challenging.

Final Diagnosis

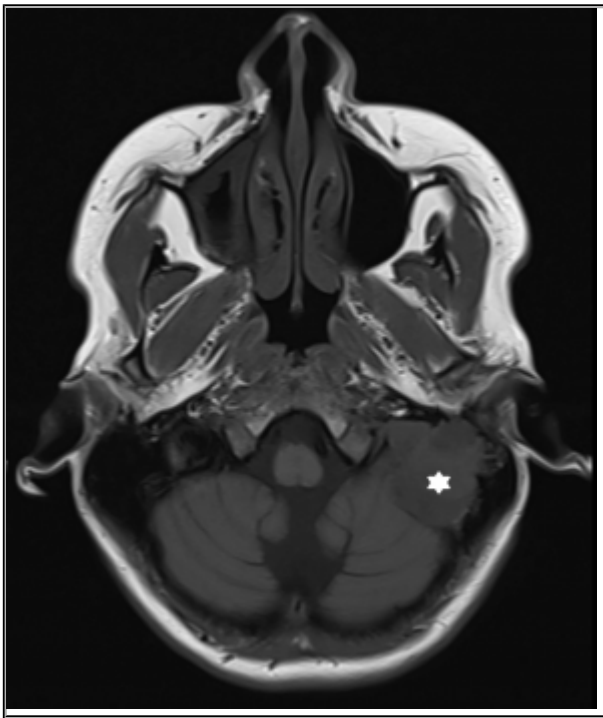
Chondromyxoid fibroma of the mastoid

Differential Diagnosis List

Endolymphatic sac tumor, Chondroma and chondrosarcoma, Facial nerve schwannoma

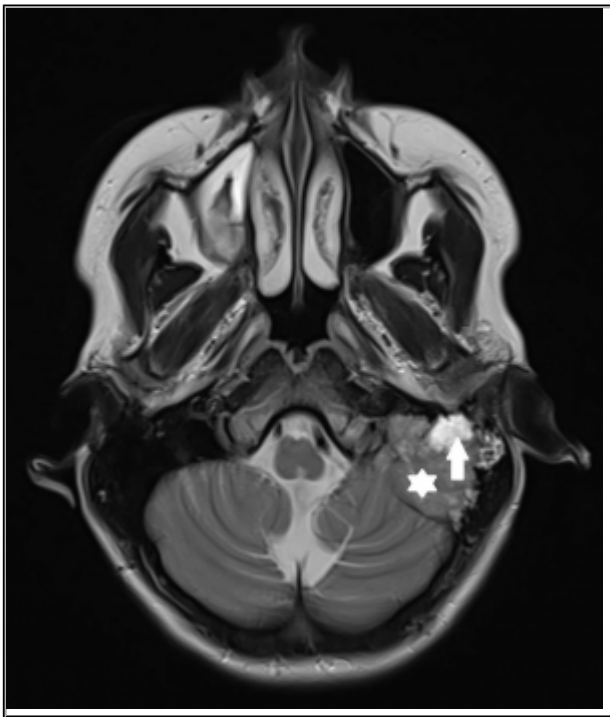
Figures

Figure 1 Axial T1-WI (A) and T2-WI (B)



Axial T1-weighted image (T1-WI) shows a space-occupying lesion of low signal intensity (SI) in the mastoid (asterisk).

Area of Interest: Head and neck;
Imaging Technique: MR;
Procedure: Education;
Special Focus: Tissue characterisation;

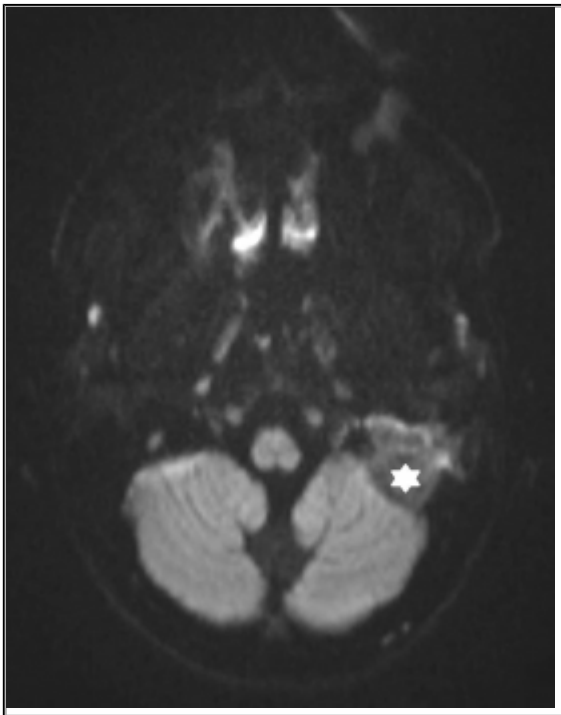


Axial T2-weighted image (T2-WI) shows a space-occupying lesion of heterogeneous

intermediate to high SI in the mastoid (asterisk).

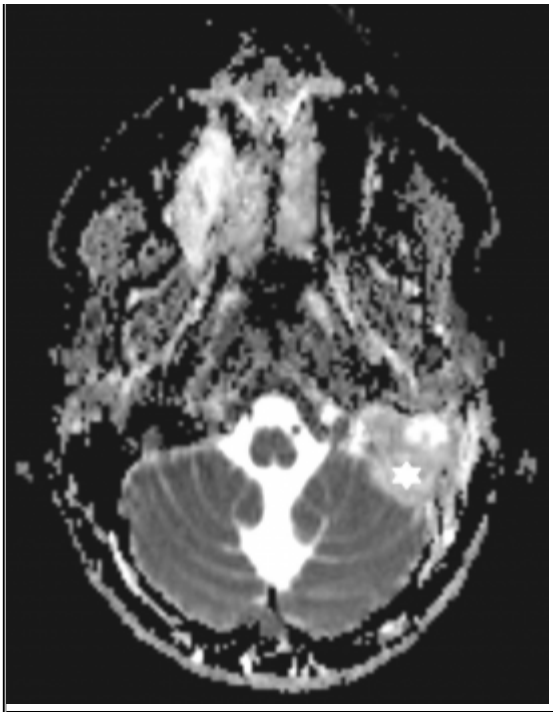
Area of Interest: Head and neck;
Imaging Technique: MR;
Procedure: Education;
Special Focus: Tissue characterisation;

Figure 2 Axial DWI (A) and ADC maps (B)



Axial diffusion weighted image (DWI) shows no significant diffusion restriction (asterisk).

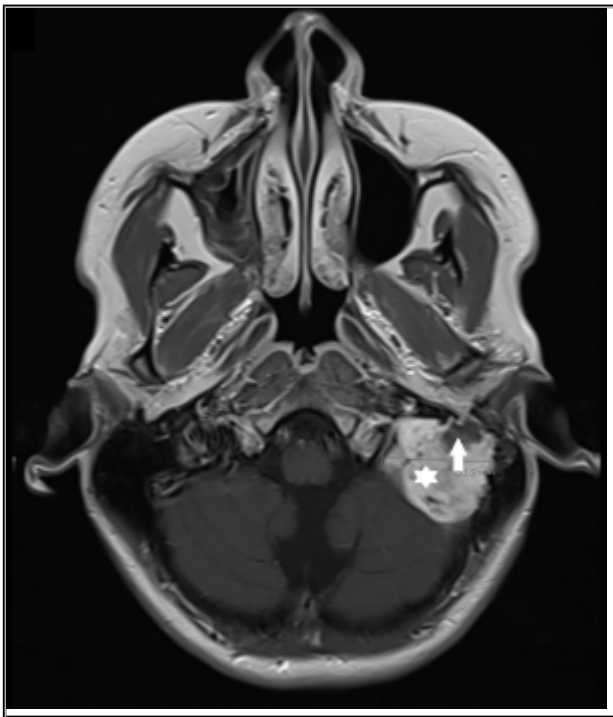
Area of Interest: Head and neck;
Imaging Technique: MR;
Procedure: Education;
Special Focus: Tissue characterisation;



Axial apparent diffusion coefficient (ADC) map shows no significant diffusion restriction (asterisk)

Area of Interest: Head and neck;
Imaging Technique: MR;
Procedure: Education;
Special Focus: Tissue characterisation;

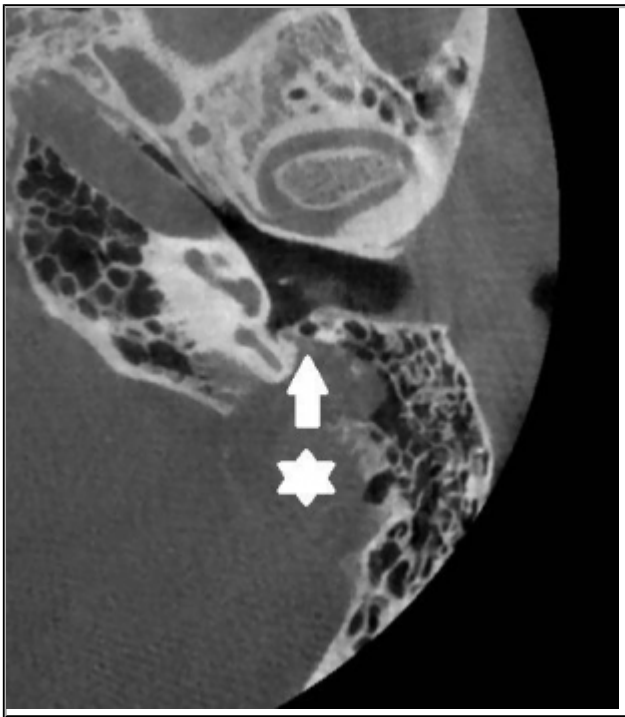
Figure 3 Axial T1-WI after intravenous administration of gadolinium-based contrast medium



On axial T1-WI after intravenous administration of gadolinium-based contrast medium, the lesion shows clear enhancement (asterisk), with exception of the anterior part, which does not enhance (arrow).

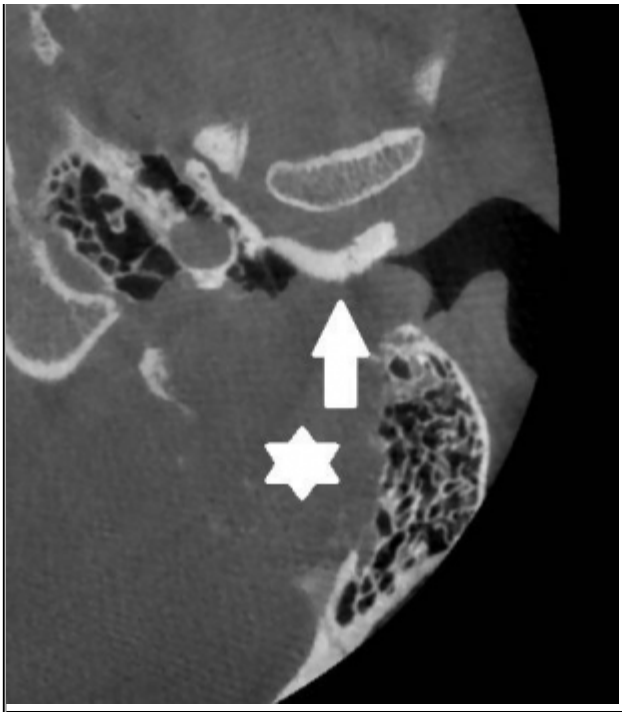
Area of Interest: Head and neck;
Imaging Technique: MR;
Procedure: Contrast agent-intravenous;
Special Focus: Tissue characterisation;

Figure 4 Axial cone-beam computed tomography of the left mastoid



Axial cone-beam computed tomography confirms the destructive osteolytic space-occupying lesion in the left mastoid (asterisk), with erosion of the facial canal (arrow)

Area of Interest: Head and neck;
Imaging Technique: CT;
Procedure: Education;
Special Focus: Tissue characterisation;



Axial cone-beam computed tomography confirms the destructive osteolytic space-occupying lesion in the left mastoid (asterisk), with erosion of the posterior wall of the external acoustic meatus (arrow).

Area of Interest: Head and neck;
Imaging Technique: CT;
Procedure: Education;
Special Focus: Tissue characterisation;

Figure 5 Overview of all published cases of CMF of the mastoid

AUTHOR	AGE	GENDER	SYMPTOMS AND SIGNS	CT	MRI	TREATMENT
Current case	21	F	Facial nerve palsy Narrowed EAC	Soft tissue mass eroding mastoid air cells	T1: low SI T2: intermediate to high SI T1+Gd: heterogeneous CE	Mastoidectomy
D'Andrea et al. 2017	19	F	Facial nerve palsy	Not performed	Soft tissue mass eroding mastoid air cells	Partial mastoidectomy
Gupta et al. 2012	42	M	Otalgia Hemotympanum	Soft tissue mass eroding mastoid air cells	T1: low SI T2: high SI T1+Gd: homogeneous CE	Mastoidectomy
Jaffe et al. 1958	67	M	NA	No further data found	No further data found	No further data found
Karunaiah et al. 2013	80	F	Facial nerve palsy, purulent ear discharge Swelling over right mastoid	Not performed X-ray: osteolytic tumor	Not performed	Mastoidectomy
Kitamura et al. 1989	48	M	Aural fullness, vertigo, tinnitus, mixed hearing loss	Soft tissue mass eroding mastoid air cells Intratumoral ca	T1: low SI T2: high SI	Mastoidectomy
LeMay et al. 1997	22	M	Otalgia, headache, conductive hearing loss	Soft tissue mass eroding mastoid air cells	T1: low SI T2: intermediate to high SI	Mastoidectomy
Oh et al. 2013	58	F	Conductive hearing loss Narrowed EAC	Soft tissue mass eroding mastoid air cells Intratumoral ca	T1: low SI T2: intermediate to high SI T1+Gd: heterogeneous CE	Mastoidectomy
Otto et al. 2007	58	F	Vertigo, syncope	Soft tissue mass eroding mastoid air cells	T1: intermediate SI T2: intermediate to high SI	Mastoidectomy
Patiño-Cardoba et al. 1998	26	M	Conductive hearing loss Narrowed EAC	Soft tissue mass eroding mastoid air cells	T1+Gd: heterogeneous CE	Mastoidectomy
Thompson et al. 2009	33	F	Facial nerve palsy	Soft tissue mass eroding mastoid air cells Intratumoral ca	T1: intermediate to low SI T2: high SI T1+Gd: homogeneous CE	Mastoidectomy

ca = calcifications; CE = contrast enhancement; EAC = external acoustic canal; NA = not available; SI = signal intensity

Overview of all published cases of chondromyxoid fibroma (CMF) in the mastoid

Area of Interest: Education;
Imaging Technique: RIS;
Procedure: Education;
Special Focus: Tissue characterisation;

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Citation

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1. Department of Radiology, Ghent University Hospital, Ghent University, Ghent
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3. Department of Pathology, Ghent University Hospital, Ghent University, Ghent
4. Department of Otorhinolaryngology, AZ Sint-Maarten, Duffel-Mechelen
5. Department of Radiology, Antwerp University Hospital, Antwerp University, Antwerp (2017, Aug. 1)
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URL: http://www.eurorad.org/case.php?id=14843