In patients with hypercholesterolemia, xanthoma formation is frequently seen over subcutaneous tissue of extensor surfaces and tendons that have received minor trauma or friction. Intracranial xanthomas are rare: only 16 such cases have been reported in the literature, and most of these were unilateral.\(^1\)\(^-\)\(^10\) We present a case of bilateral temporal bone xanthoma in a patient with hypercholesterolemia.

**Case Report**

**History.** This 35-year-old man presented to our hospital with bilateral, buzzing tinnitus and progressive bilateral hearing loss that had been worsening since he was 17 years old. He had undergone multiple mastoid surgeries over the years. He had received a diagnosis of hypercholesterolemia when he was 17, but treatment had been irregular. There was also a strong family history of hypercholesterolemia: 3 of his sisters suffered from it, and his mother had died of a myocardial infarction when she was 56 years of age.

**Examination.** There were operative scars over both mastoid regions with lower motor neuron seventh cranial nerve palsy over the right side. The patient had sensorineural deafness in the left ear and mixed hearing loss in the right. Xanthomas were present over both periorbital areas, Achilles tendons, and triceps tendons.

Analysis of the patient’s blood showed a raised fasting cholesterol level of 9.5 mmol/L, low-density lipoprotein level of 6.25 mmol/L, and high-density lipoprotein level of 0.73 mmol/L. Skull radiography demonstrated a hyperlucent lesion in the right and left temporal bones with osteosclerosis in the area surrounding the lesion.

A CT scan showed destruction of both the inner and outer tables of the skull base in the temporal regions bilaterally. There was a mass 6.5 cm in diameter extending from the left mastoid region upward into the left temporal lobe. A similar mass measuring 3 cm was also seen that arose from the right mastoid and extended up into the right temporal lobe. These masses showed some marginal enhancement, and hypodense areas of calcification were present within these lesions (Fig. 1). Magnetic resonance imaging revealed tumors of mixed signal intensity filling the mastoid cavity with intracranial extension into the middle and posterior cranial fossa over the left side, and a smaller intracranial extension over the right side. Both tumors had fatty components and inhomogeneous enhancement (Fig. 2).

**KEY WORDS** • hypercholesterolemia • temporal bone • xanthoma

**Abbreviations used in this paper:** CT = computed tomography; MR = magnetic resonance.
Operation and Postoperative Course. The patient underwent a left temporal craniotomy with radical mastoidectomy and tumor excision. The tumor was a hard, yellowish mass that was entirely extradural and densely adherent to the dural surface. There was destruction of temporal bone tissue with extension into the mastoid bone. Histological examination demonstrated fragments of fibrous tissue infiltrated by large numbers of foamy histiocytes that exhibited an abundance of cholesterol clefts and were surrounded by multinucleated giant cells (Fig. 3). The patient recovered well after surgery and was started on a lipid-lowering drug. At 14 months postoperatively there was no worsening of his symptoms. On imaging, the mass on the left side had minimally increased in size (~ 0.5 cm) and the lesion remnants on the right side remained unchanged (Fig. 4). The patient’s cholesterol level had decreased from 7.02 mmol/L to 5.86 mmol/L.

Discussion
Xanthoma involving the temporal bone is an extremely rare condition with only about 16 reported cases in the literature.1–10 This is only the second reported case in which the lesion was bilateral.7 The predisposing cause in this patient was hyperlipidemia. Xanthoma of the temporal bone is a benign lesion. Complete or even partial removal is effective as long as the predisposing cause is also treated.1–5

There are 2 theories concerning the formation of xantho-
mas. The first theory is that local trauma or hemorrhage causes lipids to leak from the vasculature into the surrounding tissues, where they are subsequently phagocytosed by macrophages. The lipids accumulate within these cells, creating foamy macrophages. The extracellular cholesterol crystallizes into clefts and induces an inflammatory reaction with giant cells and resultant fibrosis. The second theory is based on the view that lipotrophic factors from the blood in patients with autoimmune conditions may cause undifferentiated mesenchymal cells to undergo xanthomatous transformations through a series of inflammatory reactions, with the subsequent accumulation of phagocytic histiocytes.

Xanthomatous changes are observed as secondary changes in patients with hypercholesterolemia. Our patient had undergone 3 previous surgeries for the same problem but had had recurrences due to poor control of his hyperlipidemia. We achieved good postoperative control of hyperlipidemia with lipid-lowering drugs and proper diet.

In our patient radiological investigation was used to assess the extent of the lesion and its involvement of the adjacent structures. On T1-weighted MR imaging there were mixed hypo- and isointensities, and T2-weighted imaging demonstrated mixed hypo- and hyperintensities.

It is not possible to reach a definitive diagnosis based only on imaging. The final diagnosis is dependent on the intraoperative demonstration of a soft, yellow mass with a curdlike center surrounded by a hard mass that is adherent to the dura mater. Histopathologically, this lesion is composed of lipid-laden foamy histiocytes associated with cholesterol clefts and inflammation. It is important to investigate for the secondary cause, an underlying condition such as diabetes mellitus or hyperlipidemia, as this must al-

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**Fig. 3.** Photomicrographs. A: Cholesterol clefts surrounded by multinucleated giant cells and foamy histiocytes. B: Area predominated by foamy histiocytes. C: Foamy histiocytes. H & E, original magnification × 20 (A and B), and × 40 (C).

**Fig. 4.** Axial (left) and coronal (right) MR images obtained in the patient at 14 months of follow-up demonstrating that the mass in the left mastoid region had increased in size by 0.5 cm in diameter. The mass in the right mastoid remained unchanged. Arrows indicates lesions.
so be treated. The treatment for xanthoma is conservative surgical debulking, dietary restriction of fat and cholesterol, and pharmacological reduction of serum lipid levels.\(^3\)

With good lipid control, the lesion on the right side in our patient did not increase in size on follow-up imaging, and the debulked lesion showed only a small increase in size. However the patient will require life-long follow-up with either CT or MR imaging.

**Conclusions**

Bilateral xanthoma of the temporal bone is an extremely rare, benign condition. It is not possible to reach a definitive diagnosis based on imaging alone. Intraoperative and histopathological findings are essential in establishing a final diagnosis. The secondary cause, such as diabetes mellitus or hyperlipidemia, must also be investigated and treated. Lifelong follow-up is essential to monitor the progress of the disease.

**References**


