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Clinical Record

Facial nerve decompression via middle fossa approach for hyperostosis cranialis interna: a feasible therapeutic approach

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Abstract

Hyperostosis cranialis interna is an autosomal dominant disorder characterised by endosteal hyperostosis and osteosclerosis of the skull base and calvaria, leading to compression and dysfunction of cranial nerves I, II, VII and VIII.

Case report: We report the use of bilateral surgical decompression of the internal auditory canals to treat hyperostosis cranialis interna in an eight-year-old girl presenting with bilateral facial palsy due to hyperostosis cranialis interna.

Intervention and outcome: Using a middle fossa craniotomy approach, both internal auditory canals were unroofed and cranial nerves VII and VIII were decompressed, with a one-year interval between sides. The mimic function recovered. One year post-operatively, the right and left facial sides had been restored to House–Brackmann grades I and II, respectively.

Conclusion: This is the first report of the use of surgical decompression of the internal auditory canal in a case of hyperostosis cranialis interna. Surgical decompression of the internal auditory canal is recommended therapeutically, but may also be performed prophylactically in younger patients with hyperostosis cranialis interna.

Key words: Hyperostosis; Facial Nerve; Internal Auditory Canal; Middle Cranial Fossa

Introduction

Hyperostosis cranialis interna is a disorder characterised by hyperostosis and osteosclerosis limited to the calvaria and skull base. This leads to narrowing of cranial nerve foramina and hence cranial nerve entrapment, mainly of cranial nerves I, II, VII and VIII. Smell, taste and vision impairment, facial palsy and vestibulocochlear dysfunction may occur at a young age. In addition, increased ocular and intracranial pressure can occur. Hyperostosis cranialis interna has an autosomal dominant transmission pattern and was first described in 1990.[1]

Craniofacial hyperostotic disorders can lead to dysmorphic, functional and neurological symptoms. No causal treatment modalities are known. Symptomatic treatment may consist of cranial vault decompression[2] or cranial nerve decompression. Successful vestibulocochlear and optic nerve decompression has been described in cases of craniofacial hyperostotic disorder.[3–5]

In this report, we communicate our clinical experience (including surgical technique and functional outcome) of a patient with facial paralysis due to hyperostosis cranialis interna who was treated by bilateral surgical decompression of the internal auditory canals via a middle fossa approach. To our knowledge, this is the first report of such (bilateral) treatment for this condition.

Case report

An eight-year-old girl had originally presented with sudden, paresis of the left side of the face, evolving into paralysis within two days (House–Brackmann grade VI). The referring otolaryngologist had prescribed antibiotics and prednisolone. The patient's left-sided paralysis had partially resolved after such treatment, to House–Brackmann grade III. The patient's medical history was uneventful. There was a family history of facial palsy due to hyperostosis cranialis interna (Figure 1).

The child's facial appearance was otherwise normal. The otoscopic aspect of the eardrums was normal. There was no impairment of other cranial nerves, and the ophthalmological examination was normal.

Audiometry revealed normal hearing levels. The stapedial muscle reflex was absent on the left side. Vestibular examination and brainstem audiometry were normal. Routine laboratory investigation revealed no abnormalities.

Radiological imaging by means of computed tomography (CT) was performed. Bilaterally, the middle and medial parts of the internal auditory canal were narrowed down to 2–3 mm (Figure 2a and 2b). The middle and inner ear displayed no abnormalities. The skull showed diffuse, hypodense, hyperostotic areas and a hypodense, enlarged endosteal cortex of the greater wing of the sphenoid bones and the petrous bones (Figure 3). No calvarial enlargement was seen. Magnetic resonance imaging (MRI) revealed no supplementary information.

The patient's clinical symptoms, findings and imaging results confirmed the diagnosis of hyperostosis cranialis interna. The facial nerve function did not further recover. Because of the patient's age, and the risk of progressive osteosclerosis along with facial nerve function deterioration

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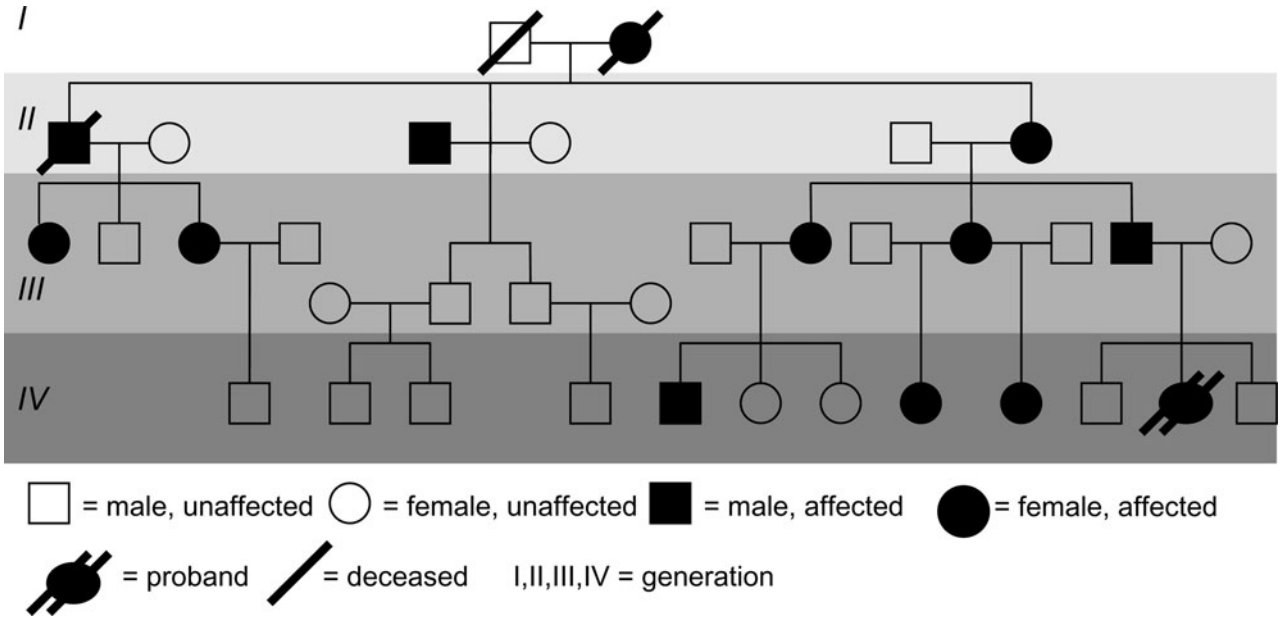

FIG. 1
Family pedigree over four generations.

with advancing age, it was decided to discuss with the parents a surgical decompression of the facial nerve via a middle fossa approach. The parents gave consent.

Ten months after the initial left-sided paralysis, the patient developed a sudden, right-sided facial palsy. Otorhinolaryngological examination revealed a partially recovered left facial palsy (House–Brackmann grade III) together with a right facial palsy (House–Brackmann grade VI, with absent stapedial muscle reflex). All other investigations remained normal. Treatment with prednisolone resulted in partial recovery of the right facial paralysis (to House–Brackmann grade IV).

The patient was scheduled for surgery firstly of the right side; it was argued that a shorter period of impairment may increase the chance of recovery. One year later, the left side was operated upon (which had in the interim remained at House–Brackmann grade III).

The surgical procedures were performed with the head fixed in a Mayfield frame. Routine monitoring of facial nerve function (using a NIM-2® XL nerve integrity monitoring system; Medtronic, Minneapolis, Minnesota, USA) and of hearing (by auditory brainstem response monitoring) was performed throughout the operation. A neuronavigation system was used (Medtronic Surgical Navigation Technologies, Louisville, Colorado, USA). Cranial to the ear, an inverted V shaped incision was made. The temporal muscle was split in an inverted T shape. The middle fossa was approached through a 5×3 cm trepanation and elevation of the dura mater. The superior petrosal sinus and the cranioposterior petrosal bone were identified. The routine landmarks for identification of the internal auditory canal could not be applied. In particular, the subarcuate eminence and the greater superior petrosal nerve could not be found. The superior semicircular canal lay buried under an abnormally thickened layer of bone which contained several large cells. The internal auditory canal was identified at the bottom of one of these cells. The consistency and aspect of the bone was normal. The internal auditory canal was pear-shaped with its narrowest point lying medially. The superior and medial parts of the internal auditory canal were drilled thin and subsequently removed.

During the first operation (on the right side), the nerves of the internal auditory canal were found to 'blossom out' into the overlying air cells. A small defect in the posterior fossa dura was repaired with a free muscle graft and fibrin glue (Tissucol®; Baxter, Deerfield, Illinois, USA). A lumbar drain was placed and left in situ for a few days.

The facial nerve function of the right facial side was House–Brackmann grade IV both pre- and post-operatively, but was completely restored (i.e. House–Brackmann grade I) one year after the operation.

The left side was operated upon one year after the right side. This procedure was similar to that on the right side. On opening the internal auditory canal, the facial nerve bulged out instantly. This normalised after administration of 4 mg intravenous dexamethasone.

One year after each operation, the right and left facial nerve function were restored to House–Brackmann grades I and II, respectively. Post-operative CT scanning revealed significantly enlarged medial internal auditory canals bilaterally (Figures 2c and 2d).

Discussion

In our patient with hyperostosis cranialis interna, marked medial internal auditory canal stenosis was present bilaterally, whereas the lateral internal auditory canal and the labyrinthine, tympanic and mastoid segments of the fallopian canal were normal. This finding justified exploration of the middle and medial internal auditory canal only. Furthermore, exploration of the unaffected lateral internal auditory canal would run the unnecessary risk of damaging the facial nerve and the labyrinth. Both surgical decompressions had the desired effect: almost complete recovery of facial nerve function. The decision to perform surgical decompression was based on the patient's early age for symptomatic hyperostosis cranialis interna, and the assumption that her osteosclerosis would worsen with age, as had happened in several relatives. Bilateral palsy had occurred in four other family members, before the advent of nerve monitoring. Despite conservative treatment, their facial nerve function had never recovered.

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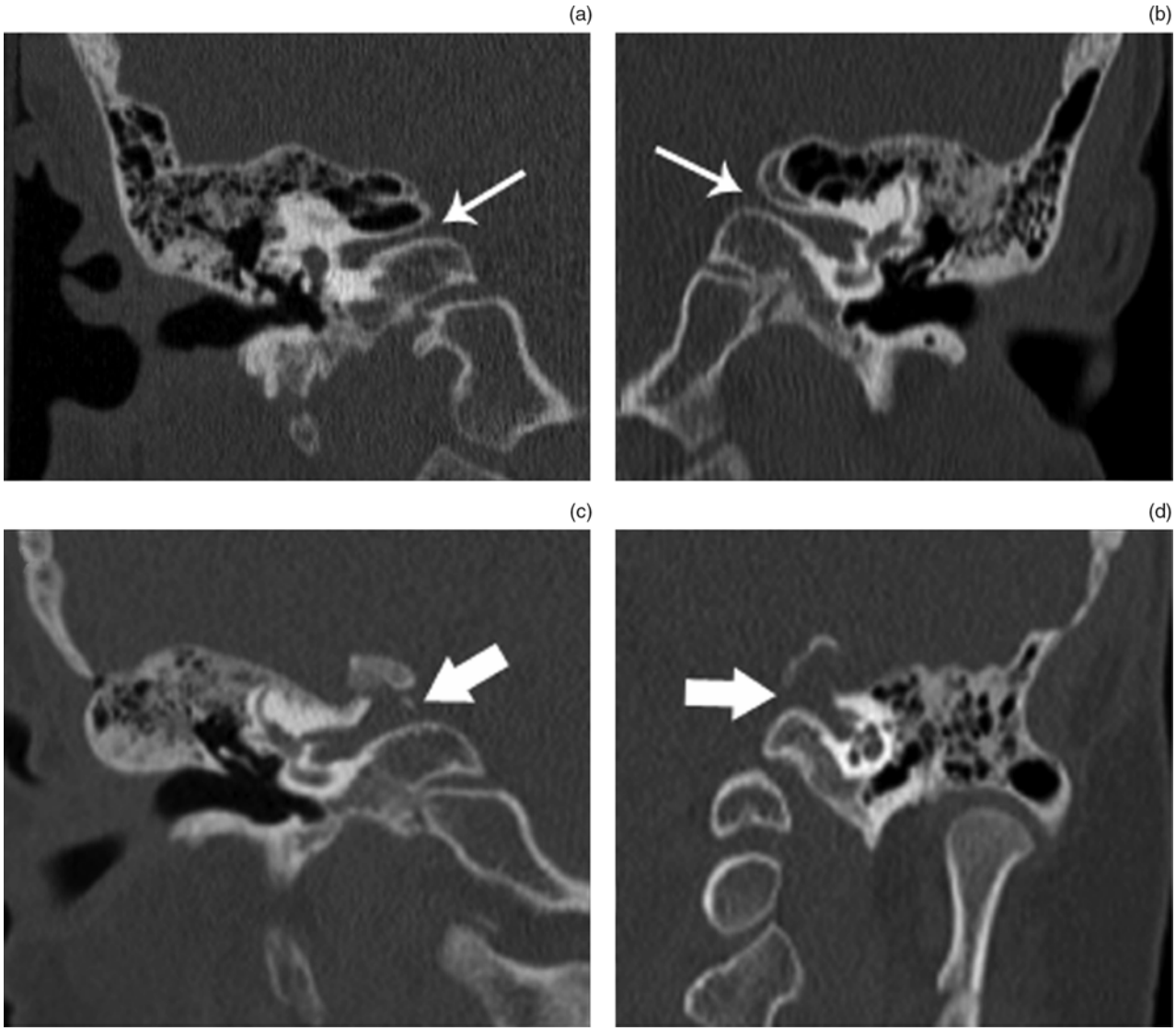

FIG. 2
Coronal computed tomography scans; the internal auditory canal is indicated by arrows. (a) Pre-operative, right side; (b) pre-operative, left side; (c) post-operative, right side; (d) post-operative, left side.

There are few published reports of cranial hyperostotic disorders treated with surgical facial nerve decompression via a middle fossa approach. Thorough analysis of the literature revealed eight reports. Hamersma mentioned four cases of meatal and labyrinthine decompression in cases of sclerosteosis performed by himself, Fisch and House.[6] Miyamoto *et al.* reported this procedure in two patients with Camurati–Engelmann disease.[7] Benecke reported successful total facial nerve decompression in an adult patient with Camurati-Engelmann disease. Moreover, Benecke reports on a middle fossa approach procedure for decompression of the internal auditory canal and labyrinthine segment in a three-year-old infant with recessive osteopetrosis.[8] Based on experiences and observations of (intracranial) complications, this author concluded that facial nerve decompression should be avoided in such severe cases. Tibesar *et al.* reported the use of internal auditory canal decompression via a middle fossa approach for treatment of sensorineural hearing loss due to vestibulocochlear nerve encroachment in a patient with Camurati–Engelmann disease.[9]

Two mechanisms can be thought of as causing facial nerve dysfunction: firstly, direct compression of the nerve by progressive hyperostotic changes of the skull base and its neuroforamina; and secondly, impairment of the perineural microvasculature. The reversibility of cranial nerve function in our patient suggests an important role for the latter mechanism, i.e. (transient) ischaemia leading to facial nerve oedema.

- **Hyperostosis cranialis interna is a disorder characterised by hyperostosis and osteosclerosis limited to the calvaria and skull base**
- **This leads to narrowing of cranial nerve foramina and hence cranial nerve entrapment, mainly of cranial nerves I, II, VII and VIII**
- **Facial nerve decompression via a middle fossa approach can be successfully performed in patients with cranial nerve entrapment due to a hyperostotic and narrowed internal auditory canal**

Hyperostosis cranialis interna is a rare disorder. However, the use of facial nerve decompression via a middle cranial

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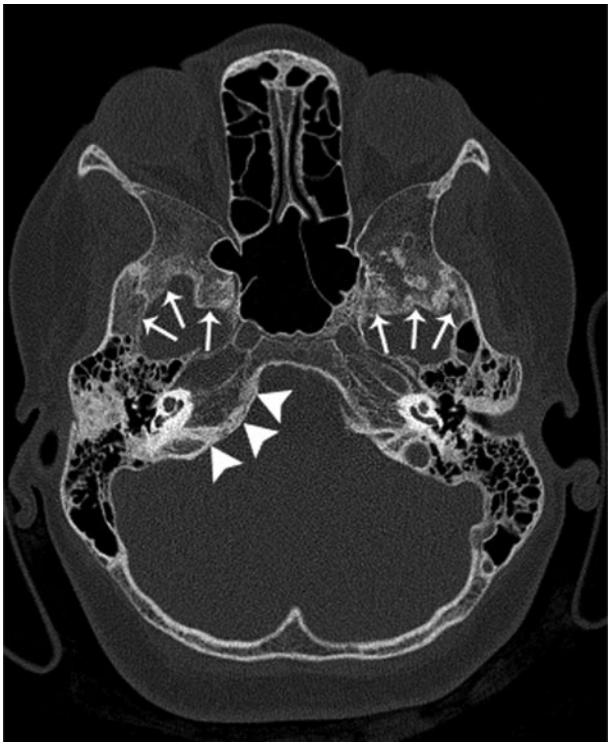

FIG. 3

Axial computed tomography scan at the level of the sphenoid bone and middle skull base, displaying diffuse abnormal findings. There is marked increased endosteal bone formation of the squama temporalis and the corpus temporalis bilaterally (arrows). Arrowheads indicate hyperostosis of the petrous bone.

fossa approach may be applicable to other hyperostotic craniofacial disorders, such as osteopetrosis, osteopathia striata, Camurati–Engelmann disease, craniodiaphyseal disease, Lenz–Majewski syndrome, Van Buchem disease, sclerosteosis and craniometaphyseal dysplasia.[10]

Conclusion

In our patient with hyperostosis cranialis interna, internal auditory canal decompression was shown to be effective in reversing facial nerve dysfunction due to encroachment of the disease. The varying manifestations of hyperostosis cranialis interna and the lack of prognostic factors indicating disease progression make it difficult to predict the clinical course of this condition. However, based on our experience with other patients with facial nerve paralysis due to hyperostosis cranialis interna, we would not expect facial nerve function to improve with conservative treatment only. Therefore, such findings call for surgical decompression at an early age. Moreover, in patients without pronounced hyperostosis and osteosclerosis of the skull, the operative landmarks are still recognisable. Therefore in young patients with hyperostosis cranialis interna, in whom progressive hyperostosis is to be expected, prophylactic surgical decompression of a narrowed internal auditory canal can be considered.

References

1 Manni JJ, Scaf JJ, Huygen PL, Cruysberg JR, Verhagen WI. Hyperostosis cranialis interna. A new hereditary syndrome with cranial-nerve entrapment. *N Engl J Med* 1990;**322**:450–4
2 Hamersma H, Gardner J, Beighton P. The natural history of sclerosteosis. *Clin Genet* 2003 Mar;**63**(3):192–7 Erratum in. *Clin Genet* 2003 Aug;**64**(2):176
3 Hellier WP, Brookes GB. Vestibular nerve dysfunction and decompression in Engelmann's disease. *J Laryngol Otol* 1996;**110**:462–5
4 Hwang JM, Kim IO, Wang KC. Complete visual recovery in osteopetrosis by early optic nerve decompression. *Pediatr Neurosurg* 2000;**33**:328–32
5 Vanier V, Miller NR, Carson BS. Bilateral visual improvement after unilateral optic canal decompression and cranial vault expansion in a patient with osteopetrosis, narrowed optic canals, and increased intracranial pressure. *J Neurol Neurosurg Psychiatry* 2000;**69**:405–6
6 Hamersma H. Total decompression of the facial nerve in osteopetrosis (marble bone disease-morbus Albers-Schonberg). *ORL J Otorhinolaryngol Relat Spec* 1974;**36**: 21–32
7 Miyamoto RT, House WF, Brackmann DE. Neurotologic manifestations of the osteopetroses. *Arch Otolaryngol* 1980;**106**:210–14
8 Benecke JE Jr. Facial nerve dysfunction in osteopetrosis. *Laryngoscope* 1993;**103**:494–7
9 Tibesar RJ, Brissett AE, Shallop JK, Driscoll CL. Internal auditory canal decompression and cochlear implantation in Camurati-Engelmann disease. *Otolaryngol Head Neck Surg* 2004;**131**:1004–6
10 Hall CM. International nosology and classification of constitutional disorders of bone. *Am J Med Genet* 2001;**79**: 376–82

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