

Bony trigeminal nerve entrapment in autosomal dominant osteopetrosis: a rare cause of secondary trigeminal neuralgia

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Case Report

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Abstract

Cranial bone disorders are a rare cause of secondary trigeminal neuralgia(TN), producing pain through progressive compression of the trigeminal nerve. We report a 46-year-old man with autosomal dominant osteopetrosis type 1 with left-sided refractory TN. Osteosclerosis of the petrous bone caused medial displacement and bony entrapment of the trigeminal nerve. Through a pretemporal extradural transcavernous approach, we unroofed the foramen rotundum and foramen ovale, drilled the petrous apex, released the calcified anterior petroclinoid fold and trigeminal fibrous ring, decompressing the nerve along its entire intracranial trajectory. The procedure was completed without complications and produced complete resolution of the TN.

Introduction

Trigeminal neuralgia (TN) is characterized by paroxysmal, electric shock–like facial pain in the distribution of the trigeminal nerve and is most often idiopathic or caused by neurovascular compression. Secondary TN arises from identifiable structural lesions such as tumors, demyelination, or skeletal abnormalities.¹ Osteopetrosis, a rare hereditary bone disorder, can cause cranial nerve compression through thickened cranial bones and progressively narrowed skull-base foramina.

Case Presentation

We present a 46-year-old male with a 4-year history of left-sided TN in the V2 and V3 distribution, refractory to medical therapies. He reported subtle bilateral hearing loss. Personal history was unremarkable except for a large head circumference. Family history revealed a father with early death after jaw surgery for bone overgrowth and one male child with macrocephaly. Neurological examination was normal. MRI demonstrated diffuse skull thickening, bilateral narrowing of the internal auditory canals, medial displacement of both trigeminal nerves due to overgrowth of the petrosal apex, and signs of intracranial hypertension with meningoceles at the lamina cribrosa. Sagittal reconstruction showed a trigeminal angle of 122°. Genetic testing confirmed ADO1 due to an LRP5 gain-of-function mutation.

The patient underwent a left-sided frontotemporal craniotomy to access the pretemporal region. Both sharp and diamond drills were used. The incision extended from the upper part of the root of the zygoma to just above the superior temporal line, within the hairline. The patient is placed supine with the head extended, rotated, and elevated above heart level. The scalp incision is made with scalpel and monopolar cautery. The cutaneous flap is reflected inferiorly and fixed with hooks. The superficial fascia of the temporalis muscle is dissected anteriorly toward the zygomatic arch, superiorly over the superior temporal line, and posteriorly, leaving a small cuff to the skull for later resuturing. The muscle is mobilized out of the subzygomatic fossa, stretched, and reflected inferiorly. Interfascial dissection of the temporalis muscle is used to protect the temporal branch of the facial nerve within the fat pad and, secondarily, for cosmetic reasons. After the bone flap is elevated, an extradural TCA is performed with focus on the lateral compartment of the CS. Due to the dense cranial bone associated with

osteopetrosis, careful drilling and bone removal were performed under high-magnification microsurgical techniques. The frontal bone and greater wing of the sphenoid are drilled with sharp and coarse diamond burrs until the periorbita is visualized. The meningo-orbital band is unroofed, coagulated, and divided. A sharp cut is made at the medial attachment of the CS roof on the lateral pillar of the anterior clinoid process, part of the lesser sphenoid wing. Progressively, from medial to lateral, the lamina propria of the temporal dura is dissected from the lateral wall of the CS using both sharp and blunt technique. The SOF, FR, and FO are identified together with their contained nerves. Hemostasis of the CS between V1 and V2 is achieved by gentle injection of tissue glue with concomitant compression of the sphenoparietal sinus to avoid retrograde embolization of the sylvian veins. The dissection is extended laterally toward the Glasscock and Kawase triangles and posteriorly toward Meckel's cave. Individual structures of the CS can be dissected by cutting tangentially around them to remove surrounding pathology.

The FR and FO were unroofed to decompress the V2 and V3 divisions of the trigeminal nerve, respectively, and to allow manipulation during surgery. Without this step, the postoperative result could be unchanged or worse owing to residual foraminal stenosis and additional injury from traction on the nerve. Hemostasis was obtained with porcine gelatin sponges and fibrin sealant. The dura over the ganglion was incised tangential to the nerve's trajectory, releasing CSF.

The Kawase rhomboid was identified and the GSPN released from the periosteal dura. The petrous bone, including the petrous apex, was drilled to create a corridor to the lateral prepontine cistern in the posterior fossa, allowing safe mobilization of the trigeminal nerve. The calcified anterior petroclinoid fold and trigeminal fibrous ring, sometimes also called the "petroclinoid ligaments", were carefully released.

The dura and tentorium were opened in a controlled fashion, exposing the cisternal segment of the trigeminal nerve and the antero-lateral pons. The edge of the petrous apex contains a sulcus for the superior petrosal sinus, which is sometimes mistaken for the boundary between the "false" and "true" petrous ridges. Before the tentorium was divided, its superior edge containing the superior petrosal sinus was coagulated and cut. Throughout the procedure, care was taken to avoid injury to the adjacent cranial nerves medially and to the petrous segment of the internal carotid artery. The trigeminal nerve was released from its arachnoid attachments and gently mobilized. Full decompression was achieved by freeing the nerve from the surrounding bony overgrowth and ligamentous constraints.

The procedure was completed without complications, achieving adequate decompression of the trigeminal nerve while preserving neurological function.

The postoperative recovery was uneventful, without neurological deficits, cerebrospinal fluid leaks, or other complications. He reported complete resolution of trigeminal neuralgia immediately after surgery. The patient remains pain-free at subsequent follow-up visits, with imaging confirming the decompressed nerve.

Discussion

Primary TN is most commonly idiopathic or caused by vascular compression of the trigeminal nerve. Secondary TN, resulting from structural lesions, is rare and requires careful evaluation. In this case, ADO1 led to trigeminal nerve traction and compression due to diffuse skull thickening. This disease is classified in the group of osteosclerotic bone diseases.²

ADO1 results from gain-of-function mutations in the LRP5 gene³, causing excessive bone formation, cortical thickening, and osteosclerosis. Clinically, patients may present with macrocephaly, frontal bossing, a flattened nasal bridge, an enlarged mandible with a decreased gonial angle, torus palatinus, and generalized skeletal hyperostosis^{4,5} and requires multidisciplinary work-up and treatment.⁵ Cranial hyperostosis causes multiple intracranial complications⁴⁻⁸. Focused on our case, it can reduce foraminal dimensions, like in this case the FR and FO, and displace the trigeminal nerve medially by petrosal bone overgrowth.⁷ Both stenotic compression and traction are predisposing to TN. The presence of bilateral narrowing of the internal auditory canal and meningoceles at the lamina cribrosa in this patient reflects the broader cranial involvement typical of ADO1.⁷

High-resolution MRI and CT imaging are essential in these patients to evaluate nerve compression, foramen dimensions, calcification of the ligamentous constraints and associated intracranial hypertension. Imaging confirmed medial displacement of the trigeminal nerve due to the large petrosal apex and foraminal stenosis.

In our patient, sagittal reconstruction demonstrated a trigeminal angle of 122° between the cisternal and cavernous part, which is significantly reduced compared to the normal physiological range of approximately 170°.⁹ This decreased angle reflects the inferior and medial displacement of the trigeminal nerve on its cisternal part.

Surgical management of osteopetrosis is technically demanding. Dense cranial bone, narrow foramina, and displaced neurovascular structures increase the risk of injury to the cranial nerves, internal carotid artery and the brain itself.

In our patient, a left frontotemporal craniotomy with a pretemporal extradural TCA allowed unroofing of the FR and FO.

Hemostasis was obtained with porcine gelatin sponges and fibrin sealant in the anteromedial and anterolateral triangles, taking care to avoid rapid, high-volume injection of fibrin sealant in the anteromedial triangle, which can provoke retrograde sylvian vein thrombosis.

Kawase drilling of the margin of the petrous bone, including the petrous apex,¹⁰ together with release of the anterior petroclinoid fold and trigeminal fibrous ring,¹¹ freed the trigeminal nerve from its constricting bony tunnel. Extended release is the most reliable way to reduce the risk of recurrence.

Reports of TN secondary to cranial osteopetrosis are extremely rare, particularly in adulthood.¹² Most published cases describe cranial nerve compression with visual and hearing loss, hydrocephalus, and

Chiari malformations^{6,8,13}, but few detail surgical intervention for TN. In the only comparable report of trigeminal neuralgia in ADO I, pain relief was achieved with a staged posterior fossa decompression and retrosigmoid microvascular decompression, relieving crowding at the root entry zone rather than the anterior bony entrapment that dominated our case.¹⁴ Our pretemporal extradural TCA instead addressed the site of compression directly to decompress the nerve along its full extracavernous course. This strategy ensures that the nerve is free from any compression from its extracranial exit site, to Meckel's cave and posteriorly to the root entry zone. This case contributes valuable insight into the management of secondary TN in patients with cranial skeletal disorders, emphasizing the need for individualized surgical strategies, with particularly in this case a more aggressive decompression to reduce the chance of recurrence at older age.

Clinicians should consider osteopetrosis in patients with TN who also have macrocephaly, craniofacial abnormalities, or family history suggestive of skeletal disorders. Multidisciplinary evaluation, including genetic counseling, high-resolution imaging, and neurosurgical planning, is essential. This approach ensures safe and effective decompression while minimizing risk in anatomically challenging patients.

Conclusion

TN secondary to cranial osteosclerosis is exceedingly rare. Recognition of underlying osteopetrosis is crucial for management planning. Surgical approaches for TN must consider altered cranial anatomy and increased risk of complications. This case highlights the importance of a multidisciplinary approach, including genetic counseling, neuroimaging, and neurosurgical planning, in patients with secondary TN and skeletal anomalies.

Abbreviations

TN
Trigeminal Neuralgia
CS
Cavernous sinus
FR
Foramen rotundum
FO
Foramen ovale
V1
Ophthalmic nerve
V2
Maxillary nerve
V3
Mandibular nerve
SOF

Superior orbital fissure
TCA
Transcavernous approach
CSF
Cerebrospinal fluid
GSPN
Greater Superficial Petrosal Nerve
LRP5
LDL Receptor–Related Protein 5
ADO1
Autosomal Dominant osteopetrosis type 1

Declarations

Ethical approval:

Not applicable.

Consent statement

The patient has consented to submission of this case report to Acta Neurochirurgica.

Competing interests:

The authors have no conflicts of interest to disclose.

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Author Contribution

SH and VV wrote the main manuscript text, prepared the figures and video. All authors reviewed the manuscript.

Availability of data and materials:

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Figures

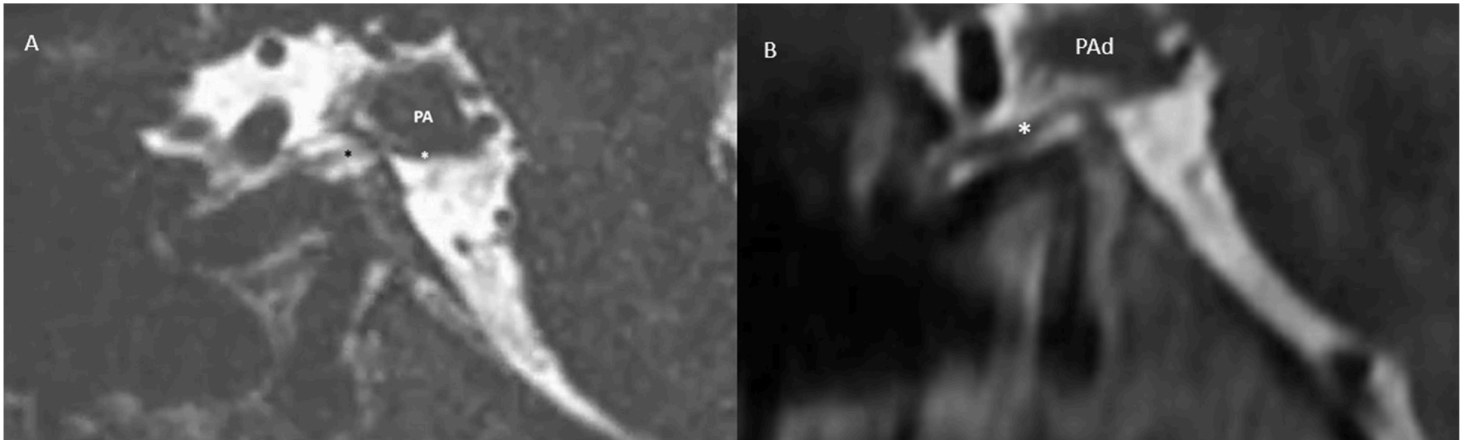


Figure 1

Picture A is showing a sagittal reconstruction of T2-weighted MRI of the trigeminal angle between the cisternal (White*) and cavernous (Black *) segments measuring 122 degrees, compared with an average of approximately 170 degrees in healthy individuals.⁹ The downward displacement is caused by the osseous protrusion of the enlarged petrous apex (PA). Picture B shows postoperative reduction of the trigeminal angle to 165 degrees(*) with the drilled petrous apex (PAd).

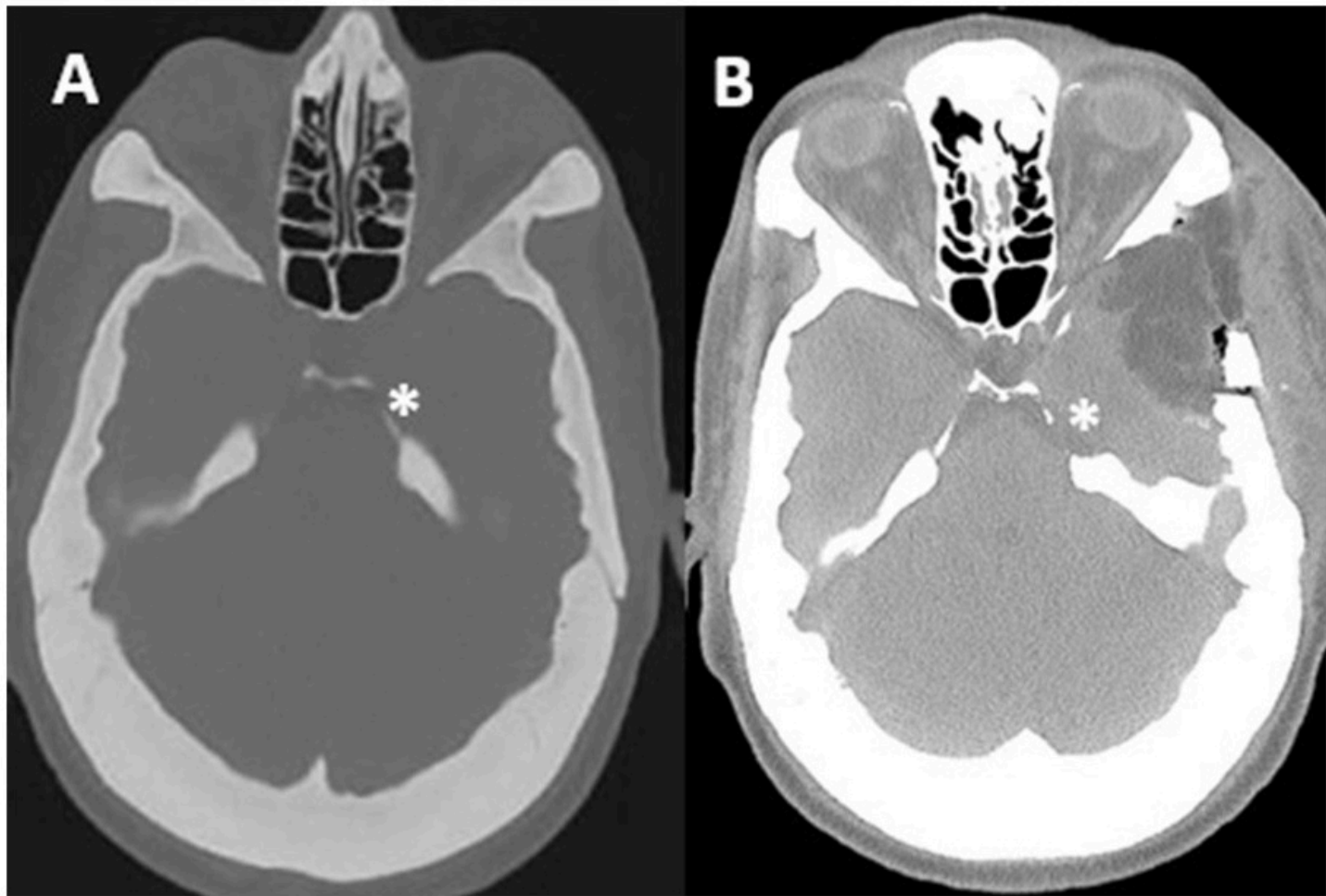


Figure 2

Bony entrapment of the trigeminal nerve under the calcified anterior petroclinoid fold and trigeminal fibrous ring (*) in picture A. Picture B shows the removal of the anterior petroclinoid fold and trigeminal fibrous ring (*) on the postoperative scan.

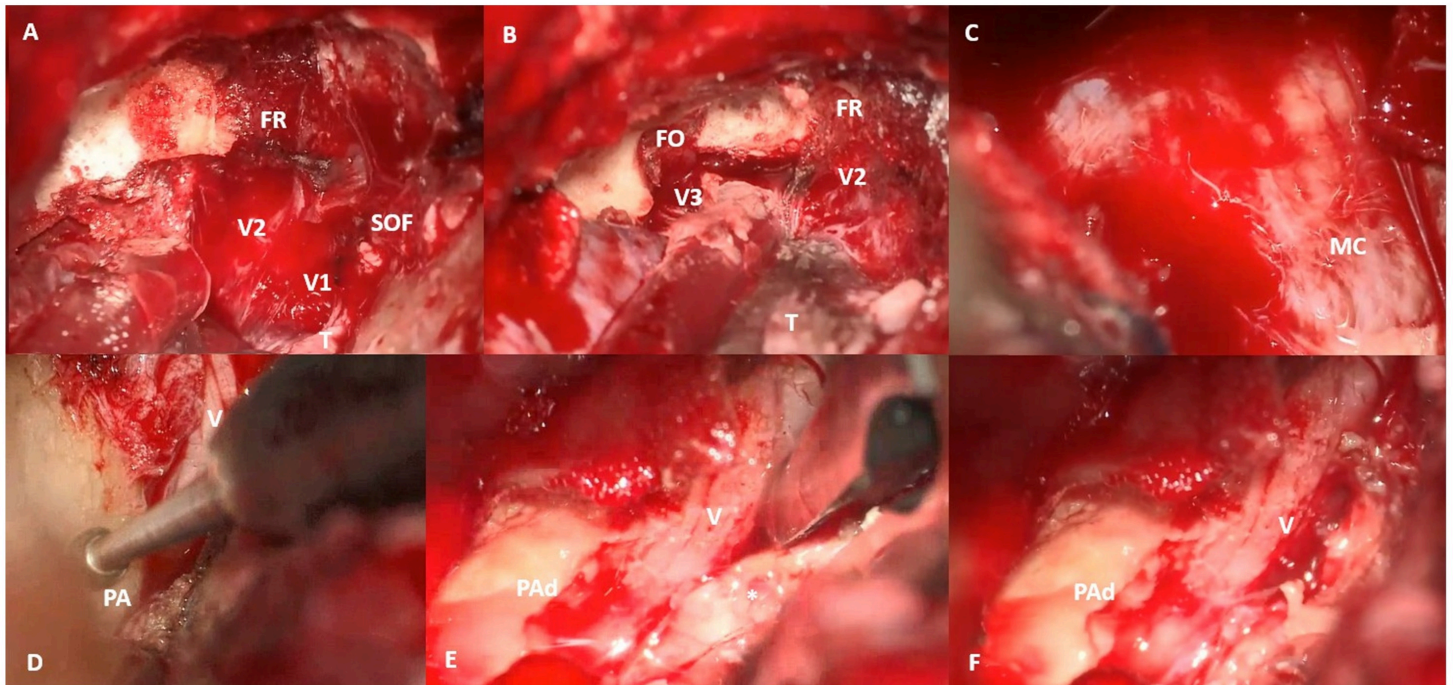


Figure 3

Surgical pictures showing in figure A the peeling of the CS. Figure B. Drilling of the roof of FR and FO. Figure C. Identification of Meckel's cave and opening of the trigeminal cistern. Figure D. Drilling of the petrosal apex lateral to the trigeminal nerve. Figure E. Removal of the calcified anterior petroclinoid fold and trigeminal fibrous ring superomedial of the trigeminal nerve. Figure F. Fully released trigeminal nerve with reduction of the trigeminal angulation and distortion. SOF – Superior Orbital Fissure, FR – Foramen Rotundum, FO – Foramen Ovale, V1 – Ophthalmic nerve, V2 – Maxillary nerve, V3 – Mandibular nerve, V – Trigeminal nerve, MC – Meckel's Cave, * Anterior petroclinoid fold and trigeminal fibrous ring, PA – Petrosal Apex, PAd – Drilled Petrosal Apex

Supplementary Files

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