Alternative option for osteogenesis imperfecta and trigeminal neuralgia

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Summary

Osteogenesis imperfecta (OI) is a bone disorder that can lead to skull base deformities such as basilar invagination, which can cause compression of cranial nerves, including the trigeminal nerve. Trigeminal neuralgia in such cases remains a challenge, given distorted anatomy and deformities. We present an alternative option, consisting in cannulation of the foramen ovale and classical percutaneous treatment. Percutaneous balloon microcompression was performed in a 28 year-old woman with OI and severe trigeminal neuralgia using computed tomography (CT) and radiographic-guided cannulation of the Gasserian ganglion without neuronavigation or stereotactic devices. The patient developed hypoesthesia on the left V1, V2 and V3 segments with good pain control. This alternative technique with a CT-guided puncture, using angiosuite without the need of any Mayfield clamp, neuronavigation systems, frame or frameless stereotactic devices can be a useful, safe and efficient alternative for patients with trigeminal neuralgia with other bone deforming diseases that severely affect the skull base.

Keywords: trigeminal neuralgia, pain, osteogenesis imperfect, percutaneous balloon compression.

Introduction

Osteogenesis imperfecta (OI), an inherited bone disorder, may produce severe disability and altered bone development, leading to multiple fractures after minimal or no trauma, thus inducing deformity.[1-3]

Often called "brittle bone disease," OI leads to various phenotypes. Mild forms can be premature or postmenopausal osteoporosis, and severe forms can lead to death in the perinatal period.[1,4]

Basilar invagination, a clinical manifestation, may cause cranial nerve compression due to odontoid process protrusion through foramen magnum into the intracranial cavity.[5,6]

The trigeminal nerve may be involved, resulting in neuralgic pain, often refractory to drug treatment. Other possible pathogeneses include arachnoid adhesions and increased vascularization in the foramen magnum area.[7-9]

Interventional treatment in such cases is difficult, mostly due to difficult access and bone fragility, besides cranial base anatomy distortion.[10]

In the past, foramen ovale cannulation was considered impossible for Gasser ganglion microcompression with conventional radiographic or tomographic-guided percutaneous approach without stereotactic or neuronavigation devices.[11]

Case report

Female patient, 28 years old, presenting OI type III, complaining of progressively worsening shooting pain with onset three years before, affecting her lower left jaw (V3 segment). Pain usually worsened while chewing, swallowing or talking and was refractory to multiple drug treatments, including carbamazepine at maximum dosage (1,200 mg daily) and pregabalin.

On physical examination, she showed multiple bone deformities, typical blue sclera, low height, marked thoracolumbar kyphoscoliosis and bilateral hearing loss, marked frontal bossing, no cranial nerve deficits, normal facial cutaneous sensation, and intact corneal reflexes.

Magnetic resonance imaging (MRI) revealed basal angle (Welcker) enlargement and clivus almost parallel to the palate, denoting platybasia. Remarkable basilar invagination was present, with odontoid process projecting 17 mm and 27 mm above the Chamberlain's line and the McGregor's line, respectively. Significant dorsal insinuation of odontoid process was found occluding pre-pontine subarachnoid space and compressing the pontine-medullary junction (Figure 1). No mass lesions or contrast enhancement over the fifth cranial nerves were seen.

Cranial-CT scan showed brachycephalic skull, with thin and irregular skull cap. Both foramina ovale showed irregular shape and reduced diameter.

The procedure was performed using angiography suite, allowing easier visualization of the foramen ovale. Under light sedation and with the patient in supine position, with slight neck extension, the X-ray intensifier was positioned on submental view to obtain the images followed by 3D CT-scan in the angiography suite. A #15 gauge cannula was inserted and positioned at the foramen ovale prior to a #4 Fogarty embolectomy catheter. The X-ray intensifier was displaced to lateral view and inflation was performed (1.0 mL of radiopaque dye). A pear-shaped image was obtained and the patient developed transitory bradycardia response. Position was confirmed with 3D CT-scan (Figure 2), balloon was deflated, process was repeated twice for 3 inflations (20 seconds/each, total 60 seconds) (Figure 3). No neuronavigation system, Mayfield clamps, frames or stereotactic devices were used.

Post-procedure, the patient reported painless hypoesthesia on left V1, V2 and V3 segments. She was discharged pain-free on the same day, with instructions to maintain the medication dosage until her first medical appointment. After one year of follow-up, the patient no longer complained of pain and ceased taking all medication.

Discussion

OI can be characterized by bone fragility secondary to reduced bone mass.[2,3] Fortunately, this clinically and genetically heterogeneous group of heritable disorders is

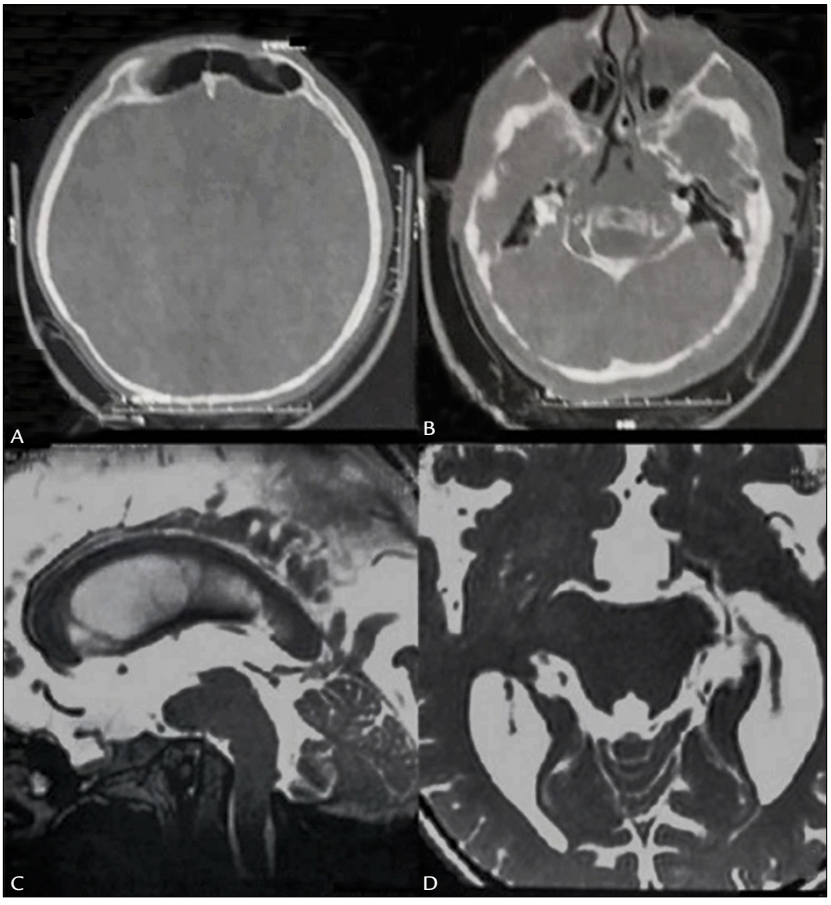

FIGURE 1 A. Wormian bones in a brachycephalic skull. B. Skull base abnormality. C. Basilar invagination obliterating the prepontine subarachnoid space, with compression of the brainstem. D. FIESTA image depicting the cisternal portion of the left trigeminal nerve.

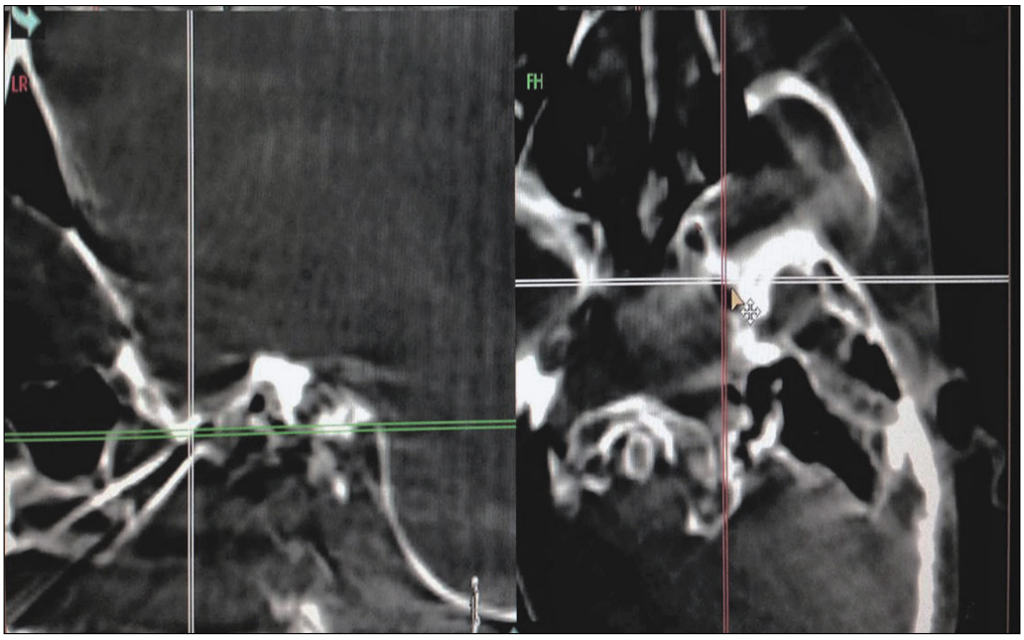

FIGURE 2 Submental view of the cannula positioned at the foramen ovale.

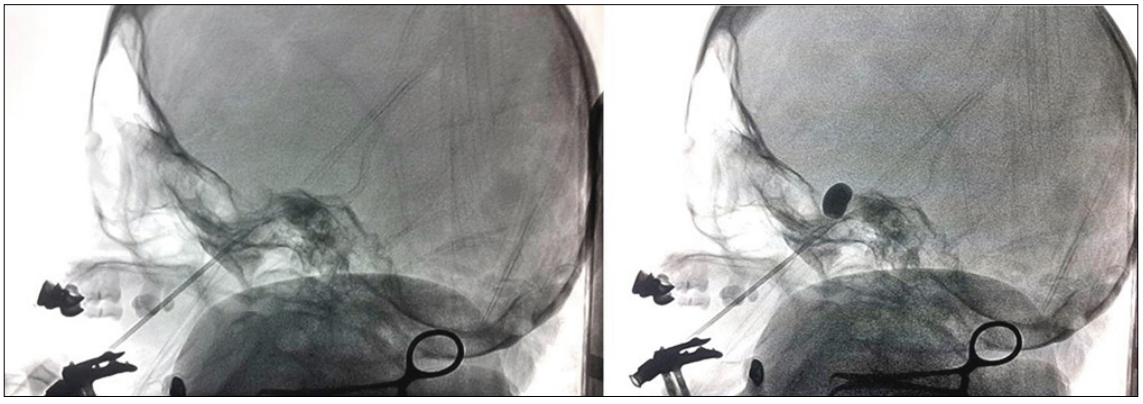

FIGURE 3 Intensifier control. Lateral view showing the pear-shaped aspect of the balloon.

mild in most cases and causes minimal deformities in adults. Skeletal manifestations may include thinned calvarium and excessive wormian bone formation.[4]

Basilar invagination is a rare complication of multiple generalized bone diseases, and may cause severe disability. The upward displacement of the basilar and condylar portions of the occipital bone may cause the foramen magnum to fold inward with subsequent translocation of the upper cervical spine into the brainstem.[7,9] This condition can result in bulbar dysfunction and myelopathy with lower cranial nerve palsies due to stretching.

Few cases in the literature described basilar invagination associated with trigeminal neuralgia.[3,8,12] This condition, due to the complexity of anatomical deformities, hinders treatment options. Microsurgical decompression may be considered as a definitive and long-term solution, but it carries a high risk, since substantial bone deformity caused by the disease may worsen after bone removal for craniotomy.

The foramen ovale approach is an alternative option, although it can prove challenging or even impossible. Normal anatomical relations are distorted by bone deformity and the lower surface of the foramen ovale may be posteromedially displaced, thus making the conventional route for percutaneous approaches virtually impossible to use.[5,7,8] The authors found that Mayfield clamps, necessary in the previous alternatives, also imply high complication risk given the potential fragmentation of wormian bones and deformity worsening.

Conclusion

In cases of OI, cannulation of the foramen ovale and classical percutaneous treatment can be a useful, safe and efficient alternative using CT-guided puncture for patients with trigeminal neuralgia and other bone deforming diseases that severely affect the skull base.

Conflict of interest

The authors declare no conflict of interest.

Resumo

Opção alternativa em tratamento da neuralgia do trigêmeo com osteogênese imperfeita

Osteogênese imperfeita (OI) é uma doença óssea que pode levar a deformidades de base de crânio, como invaginação basilar que pode provocar compressão de nervo craniano, incluindo o nervo trigêmeo. Nestes casos, a neuralgia do trigêmeo permanece como um desafio, pela anatomia distorcida e pelas deformidades. Apresentamos uma alternativa que consiste na canulação do forame oval e no tratamento percutâneo clássico. A microcompressão percutânea por balão foi realizada em uma paciente de 28 anos apresentando OI e grave neuralgia do trigêmeo, sendo realizadas tomografia computadorizada (CT) e canulação guiadas do gânglio gasseriano sem neuronavegação ou dispositivos estereotáxicos. A paciente apresentou hipoestesia à esquerda dos segmentos V1, V2 e V3, com bom controle da dor. Essa técnica alternativa com punção orientada por CT utilizando o *angiosuite* sem a necessidade de qualquer grampo de Mayfield, sistemas de neuronavegação, ou dispositivos com ou sem arcos estereotáxicos, pode ser uma opção útil, segura e eficiente para pacientes com neuralgia do trigêmeo cursando com outras doenças deformativas que afetem a base craniana de modo grave.

Palavras-chave: neuralgia do trigêmeo, dor, osteogenesis imperfeita, compressão percutânea por balão.

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