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Trigeminal Neuralgia Associated with Achondroplasia. Case Report with Literature Review

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Summary

A 59-year-old male with a history of 2 years of typical trigeminal neuralgia manifested the characteristics of achondroplasia. X-ray and magnetic resonance imaging demonstrated basilar impression, deformity of the posterior fossa with marked asymmetry of petrous bone and compression of pons and trigeminal nerve by the left vertebral artery and anterior inferior cerebellar artery. Microvascular decompression was performed through a suboccipital craniectomy. The neuralgia disappeared soon after surgery and remains completely resolved until today. This is the first reported case of trigeminal neuralgia in a patient with achondroplasia. The deformity of the skull base was considered to influence the development of the trigeminal neuralgia.

Keywords: Achondroplasia; basilar impression; trigeminal neuralgia.

Introduction

Achondroplasia is a hereditary, congenital disturbance of epiphyseal chondroblastic growth and maturation with the characteristic appearance of disproportionate short stature. Achondroplasia occasionally occurs in association with basilar impression [5, 8].

We describe a case of achondroplasia causing basilar impression and asymmetry of the petrous bone, resulting in trigeminal neuralgia. Reviewing the literatures, we found this to be the first report of a patient with trigeminal neuralgia in combination with achondroplasia.

Case Report

A 59-year-old male came to our hospital on April 1999, complaining of severe lancinating pain in the face, exclusively confined to the area of the left mandibular and infra-orbital nerves. Facial movement, such as talking, chewing and eating, aggravated the pain. Trigger zone was located in the left lower buccal region. Medical treatment with carbamazepine was initially successful, but the patient later developed refractory pain and was unable to tolerate the side-effects of medication.

On admission, he measured 131.9 cm in height (less than the third centile) with short limbs, bowed legs, normal trunk and normal vault. Neurological examination showed no abnormalities other than left trigeminal neuralgia. There was no loss of the strength of the muscles of mastication. He had a 10-year history of hypertension.

An X-ray film of the skull in the lateral view showed basilar impression (Fig. 1A). The posterior arch of the atlas was attached to the occipital squama. An X-ray film in the anteroposterior view disclosed a marked elevation of the left petrous bone (Fig. 1B).

Magnetic resonance (MR) imaging demonstrated the deformity of the posterior fossa, upward displacement of brainstem, elongated medulla and ventral compression of the pons by the left vertebral artery (Fig. 2A, B). MR angiography and constructive interference in steady-state (CISS) image showed marked elongation of the left vertebral artery compressing the root entry zone of left trigeminal nerve (Fig. 2C, D).

A left suboccipital craniectomy was performed in the right lateral position under general endotracheal anesthesia on Sep. 1, 1999. Under the operating microscope, the posterior fossa was extremely narrow. The trigeminal nerve was compressed and stretched by the tortuous vertebral artery and the anterior inferior cerebellar artery (Fig. 3A, B). Small pieces of Teflon felt were placed between the trigeminal nerve and the arteries. The postoperative course was uneventful and his trigeminal neuralgia completely resolved. The duration of relief has lasted for 18 months to date.

Discussion

Trigeminal neuralgia associated with deformity at the skull base has been reported in several cases [3, 6, 7]. In such cases, so-called crowding of the posterior fossa is thought to be responsible for the cause of the neuralgia. Basilar impression, the commonest craniocervical deformity, causes narrowing of the posterior fossa due to the upward displacement of basilar and condylar portions of the occipital bone.

Neurological deficits encountered in patients with basilar impression are varied. The most frequent symptom of basilar impression is headache and it is

Fig. 1. (A) Lateral skull radiograph showing basilar impression and atlanto-occipital assimilation. The odontoid process was found to be lying far above Chamberlain's line (*--------*), which is defined as a straight line drawn from the posterior margin of the hard palate to the posterior margin of the foramen magnum. (B) Anteroposterior radiograph revealing the marked elevation of the left petrous bone (arrowhead)

characteristically located in the neck and occipital region [3]. Involvement of the cranial nerves has been described but seems to be relatively uncommon. Among them, the trigeminal nerve is commonly involved [3, 4]. De Barros *et al.* reported that 30% to 50% of patients with basilar impression showed sensory symptoms of trigeminal nerve [1]. Gardner and Dohn found a high incidence of basilar impression in elderly women with trigeminal neuralgia [2]. Contrary to these reports, Obrador *et al.* stated that the presence of trigeminal neuralgia was less than 1% in 200 reported cases of basilar impression and other occipitocervical malformations [7].

Achondroplasia is an autosomal dominant disease. Major features are disproportionate short stature, i.e. limbs more severely shortened than trunk, macrocephaly, occasionally hydrocephaly, small foramen magnum, depressed nasal bridge, short hands and feet, limited elbow extension, caudal narrowing of spinal canal, cervical or lumbar spinal nerve compression [9]. Occasionally achondroplasia occurs in association with basilar impression [5, 8]. A case reported by Luyendijk *et al.* showed progressive tertaparesis due to medullary compression resulting from basilar impression and upper cervical stenosis [5]. Another case reported by Uematsu *et al.* (patient 2) presented with urinary incontinence and progressive claudication [8]. No patients with trigeminal neuralgia in combination with achondroplasia have been described in the literature.

In our case, trigeminal neuralgia was caused by vascular compression of the root entry zone. We presume that the marked deformity of the skull base has considerably influenced the development of the trigeminal neuralgia. Obrador *et al.* reported a case with trigeminal neuralgia associated with striking asymmetry of the petrous bone due to unilateral basilar impression [7]. They concluded that severe angulation and stretching of the trigeminal root caused by elevation of the petrous bone induced the neuralgia. As the patient underwent a partial root section via a subtemporal extradural approach, the possibility of vascular compression at the root entry zone is unknown. Microsurgery in our patient disclosed that the left trigeminal, facial and vestibulocochlear nerves were markedly displaced upward. The tortuous vertebrobasilar artery protruded superolaterally and pushed into the lateral part of pons. The left vertebral and anterior inferior cerebellar artery strongly compressed the trigeminal nerve root at its very narrow prepontine portion.

In conclusion, we consider that these anatomical

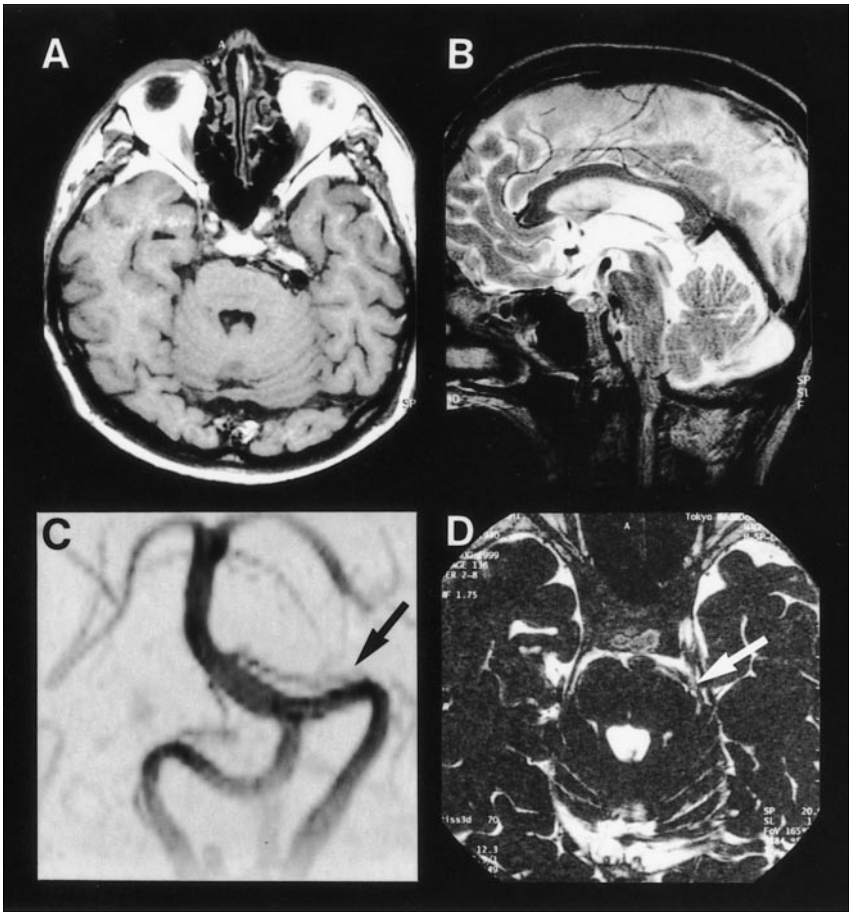

Fig. 2. Magnetic resonance images showing the upward displacement of brainstem and ventral compression of the pons by the left vertebral artery on the T1 weighted axial (A) and T2 weighted sagittal (B) images. Magnetic resonance angiography (C) and constructive interference in steady-state image (D) demonstrating marked elongation of the left vertebral artery compressing the root entry zone of the left trigeminal nerve (arrows)

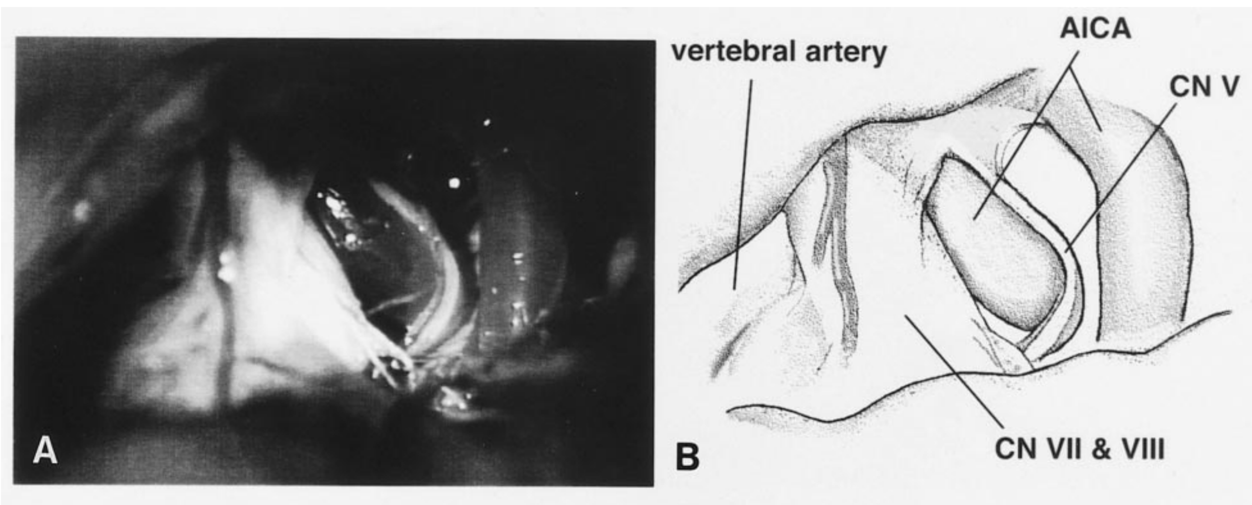

Fig. 3. Intra-operative photograph (left) and drawing of the left suboccipital retrosigmoid approach before microvascular decompression. The trigeminal nerve *(CN V)* is seen elevated superiorly by the vertebral artery and the anterior inferior cerebellar artery *(AICA)*. The facial and vestibulocochlear nerves *(CN VII & VIII)* are also displaced upward, hiding the trigeminal root entry zone

deformities at the skull base had played a causative role in the development of trigeminal neuralgia in this case as well as the tortuous vertebrobasilar system.

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Comment

This report is important because it points out that, besides the neurovascular conflict(s), morphology of the skull and lesions affecting the bony walls of posterior and middle fossae can play an important role in the pathogenis of so-called idiopathic trigéminal neuralgia

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