Orofacial Pain as the Sole Manifestation of Syringobulbia-Syringomyelia Associated with Arnold-Chiari Malformation

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This is a case report of a male patient who presented with orofacial pain for a year as the only manifestation of syringobulbia-syringomyelia associated with Arnold-Chiari malformation. This article places emphasis on the clinical presentation and possible differential diagnoses. The pain was continuous and affected the left side of the face. It was exacerbated by coughing and physical effort, possibly as a consequence of an increase in intracranial pressure. Paroxysmal pain crises developed over this background of continuous pain, compatible with neurogenic trigeminal pain of the left second branch, together with pain episodes similar to cluster headache on the same side. The symptoms were resolved following neurosurgical management with amplification of the foramen magnum.

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Syringomyelia, or cavitation within the spinal cord, represents a chronic and progressive disorder primarily involving the spinal cord. It is clinically characterized by loss of pain and temperature sensitivity, with relative preservation of touch and proprioception, amyotrophy, and paraparesis. Although the exact incidence of syringomyelia is not known, it is considered to be rare. The disease usually appears in the third or fourth decade of life. Syringomyelia usually progresses slowly, with a course extending over many years.[1]

The syringomyelic cavity, or syrinx, sometimes extends superiorly into the medulla, producing syringobulbia syndrome. This is evidenced by dysphagia, pharyngeal and palatal weakness, asymmetric weakness and atrophy of the tongue, sensory loss primarily affecting pain and temperature sense in the distribution of the trigeminal nerve, and nystagmus. Features of Arnold-Chiari malformation, such as displacement of the cerebellar tonsils into the cervical canal, are often identified.[1,2]

Described here is the case of a patient with a 1-year history of orofacial pain as the only manifestation of syringobulbia-syringomyelia associated with Arnold-Chiari malformation. Emphasis is placed on the clinical presentation and the possible differential diagnoses.

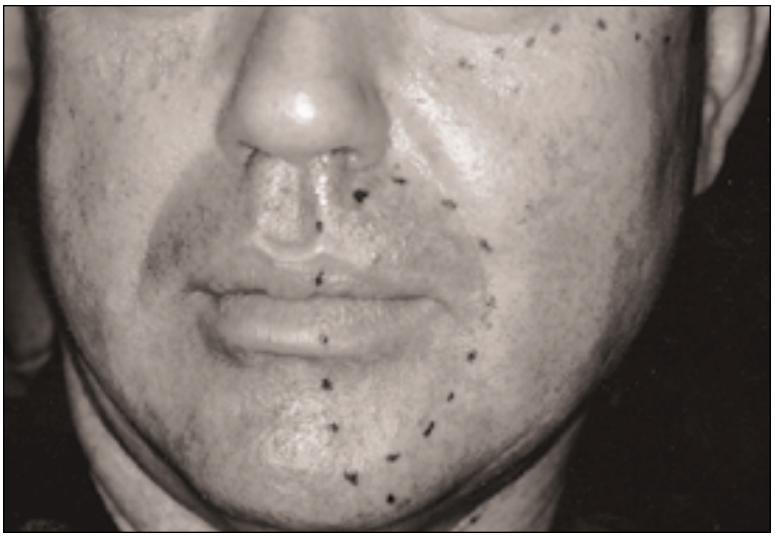

Fig 1 Clinical image of the patient, showing the zones referred to as painful.

Case Report

A 45-year-old male presented with a history of headaches beginning at age 20 and lower left hemifacial pain for the past year. The latter was continuous and moderately intense and appeared when the patient coughed or exerted physical effort. The patient also reported strange sensations in the lower left half of the face, with numbing in the left side of the neck. The pain was initially attributed to periapical pathology of a maxillary left premolar. The tooth was first subjected to endodontic treatment and then removed, although the pain persisted. A posterior cranial computed tomography (CT) study proved normal. Trigeminal neuralgia was diagnosed and treatment was provided in the form of carbamazepine 600 mg/day and ergotamine 3 mg/day. However, the patient continued to suffer pain on coughing or with physical straining, and subsequently abandoned the medication.

Over the past 3 months the pain had increased in intensity, extending from the perioral zone to the entire lower left side of the face (Fig 1) and radiating to the left side of the neck. In addition, pain crises lasting only a few seconds appeared several times daily in the territory of the left second trigeminal branch. In addition, the patient described 2 to 3 weekly episodes of pain lasting for half an hour each and extending from the premolar zone to the entire left side of the face; these were associated with tearing of the left eye and nasal congestion of the same side. The intensity of pain did not increase with either mastication or speech.

Intraoral exploration yielded no significant findings. Pain was induced by palpation of the masticatory muscles on the left side of the face, although there was no pain to palpation of the left temporomandibular joint and the intraoral structures appeared normal. A neurologic exploration of the cranial nerves was unremarkable. Trigeminal sensory exploration revealed no hypoesthesia.

Neurogenic trigeminal pain was diagnosed, with associated cluster headache–like vascular pain crises, and treatment was started in the form of carbamazepine 600 mg/day and ergotamine 3 mg/day. The overlying brief pain crises and episodes of facial pain subsequently improved, although the continuous background pain persisted.

Magnetic resonance imaging (MRI) of the temporomandibular joint revealed no unusual findings. Masticatory pain secondary to vascular and neurogenic pain was diagnosed, a stabilization occlusal splint was placed, and physiotherapy exercises were programmed. The carbamazepine and ergotamine treatment was continued and supplemented with amitriptyline (75 mg/day).

One month later, the patient had failed to improve; the continuous, lower hemifacial pain persisted and increased with coughing and physical effort. Trigeminal sensory exploration revealed thermo-analgesic dissociation (ie, a dissociation between pain and temperature, and touch and proprioception) in the lower left half of the face, predominantly in the perioral zone, with a lack of sensitivity to heat and pain. An MRI study of the head and neck revealed syringomyelia at the

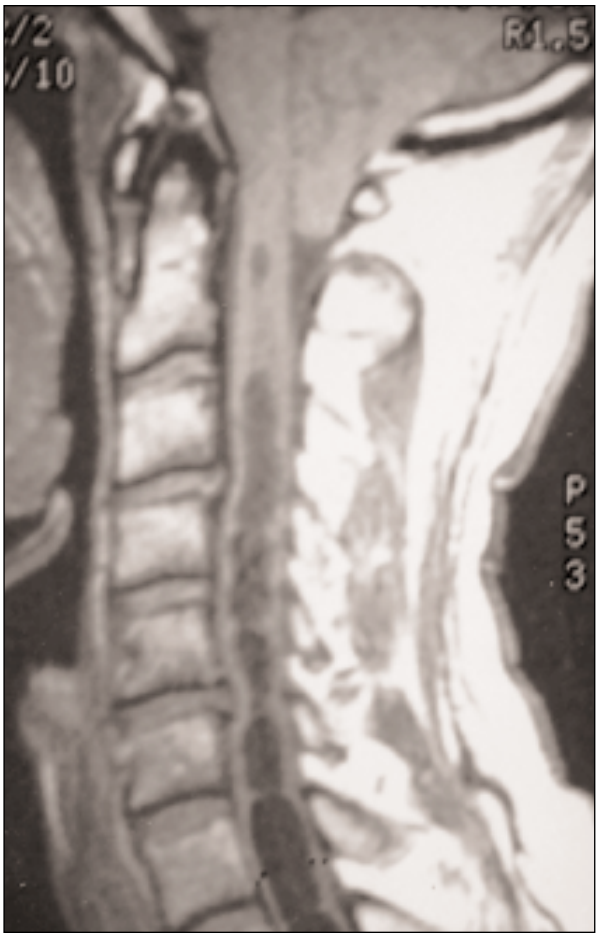

Fig 2 Brain MRI showing the characteristic cerebellar tonsil herniation of Arnold-Chiari malformation and the presence of syringomyelic cavities in the cervical medulla.

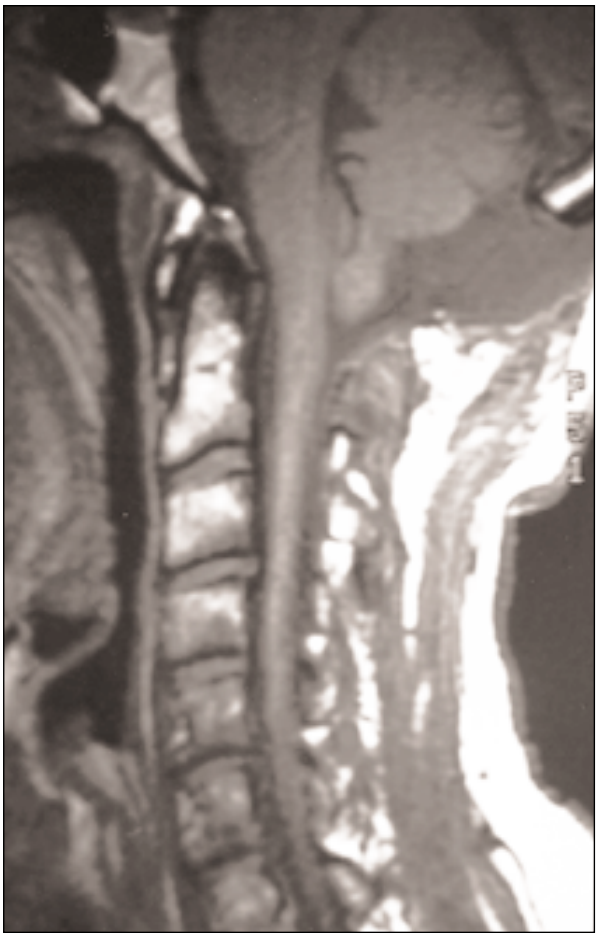

Fig 3 Postoperative MRI showing amplification of the foramen magnum and disappearance of the dilated ependymal cavities.

cervical level, syringobulbia, and Arnold-Chiari malformation with tonsillar ectopia (Fig 2).

The patient was subjected to surgical decompression of the foramen magnum combined with transverse microincisions in the outer layer of the dura; the global symptoms subsequently resolved. Follow-up MRI showed improvement of the syringomyelia (Fig 3). Two years later, the patient remained asymptomatic and was receiving no pharmacologic treatment.

Discussion

The symptomatic manifestation of syringomyelia depends primarily on the precise location of the lesion within the neuraxis. There may be signs and symptoms of injury to the cerebellum, medulla, and the lower cranial nerves, with or without evidence of increased intracranial pressure.[2] The Arnold-Chiari malformation is a congenital anomaly of the hindbrain characterized by a downward elongation of the brain stem and cerebellum into the cervical portion of the spinal cord.[3] Diseases of the cranio-vertebral junction, eg, syringomyelia-syringobulbia and tonsillar descent, can produce mechanical or hydrodynamic head and neck pain.[3] Our patient suffered continuous hemifacial pain that increased in a characteristic manner, possibly in relation to a rise in intracranial pressure caused by coughing or physical effort. The neurogenic-type pain crises that affected the second trigeminal branch could be attributed to changes in intracranial pressure affecting the trigeminal spinal tract that runs rostrocaudally along the dorsolateral margin of the medulla, and possibly the trigeminal root.[4] This type of pain responded favorably to carbamazepine.

Vegetative disorders are common in syringomyelia and include vasomotor and secretory alterations and especially Horner's sign on the side of the cavitation. There have been reports of retro-orbital pain mimicking a cluster headache attack or paroxysmal hemicrania in the cervical cord, and dorsal medullary infarction presenting with retro-orbital pain.[5,6] Our patient suffered cluster headache–like attacks with vegetative manifestations that responded well to ergotamine.

The syringomyelic cavity, or syrinx, is most commonly found in the lower cervical region, particularly at the base of the posterior horn and extending into the central gray matter and anterior commissure of the cord. By virtue of its location, the cavitation interrupts the decussating spinothalamic fibers that mediate pain and temperature sensibility, resulting in loss of these sensations. This pattern of loss of cutaneous sensitivity with preservation of posterior column sensory modalities is commonly referred to as *dissociated sensory loss*. Pain and temperature sensations are typically impaired in the arm on the involved side, although sometimes both arms are affected, or a shawl-like distribution is observed across the shoulders and upper torso anteriorly and posteriorly. Extension of the lesion into the anterior horns with resultant loss of motor neurons in turn causes amyotrophy that begins in the small muscles of the hands, ascends to the forearms, and ultimately affects the shoulder girdle muscles.[1]

When the brain stem is affected (syringobulbia), ependymal dilatation first affects the lowermost or most caudal regions of the trigeminal nuclei (particularly the caudalis and interpolaris subnuclear regions), which receive afferent inputs related to pain and temperature sensations. The higher or more rostral regions (eg, the principal and mesencephalic nuclei), which receive fine tactile or proprioceptive sensory inputs, remain unaffected; this in turn accounts for the thermo-analgesic dissociation observed in patients with syringomyelia.[1] Our patient reported a numbing sensation that extended from the perioral region to the rest of the face. The sensory alterations detected by the clinical exploration manifested as loss of heat and pain sensation, although fine tactile perception and proprioception remained intact.

The regions within subnucleus caudalis corresponding to the zones concentrically distributed around the mouth and nose are arranged from rostral to caudal; this explains the onion peel–like progression of anesthesia in ascending syringobulbia.[7] As occurred in our patient, the sensory alterations followed the somatotopic organization of the trigeminal subnucleus caudalis, ie, from the lips backward along the rest of the face.

There is no specific therapy for syringomyelia associated with Chiari type I malformation. Surgical management is widely advocated. In this sense, posterior fossa decompression, with removal of the posterior rim of the foramen magnum and of the arches of the atlas and axis (with reduction of an associated Arnold-Chiari malformation), sometimes appears helpful.[1] Bony foramen magnum decompression combined with transverse microincisions of the outer layer of the dura mater[8] may provide effective relief of symptoms.

This case report serves to remind the clinician of the many different sources of orofacial pain. Although the patient's chief complaint was initially thought to be dentally related, dental therapy failed to resolve the pain. Continued investigation finally revealed the true origin of the pain, allowing successful therapy.

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