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Radiology in focus

Primary brain stem tethering: a rare cause of geniculate neuralgia

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Abstract
This rare case of brain stem tethering presented with chronic and progressive geniculate neuralgia. In view of the fact that an occipital subcutaneous lipoma had been resected in childhood, it probably concerned a primary tethering, fitting in with an occult occipital dysraphism.

Magnetic resonance imaging (MRI) clearly demonstrated an underlying tethering, causing a distortion of the brain stem. Consequently, this led to the hypothesis that the geniculate neuralgia could be explained by traction on the lower cranial nerves secondary to the brain stem displacement. Untethering resulted in a considerable decrease of the neuralgia.

MRI proved to be essential in the diagnosis and treatment of this unusual case.

Key words: Geniculate ganglion; Neuralgia

Introduction

Geniculate neuralgia is a rare cause of otalgia described by Ramsey Hunt (Hunt, 1907). In its most typical form, the pain is severe, paroxysmal and localized deep in the ear.

A chronic geniculate neuralgia can also irradiate to surrounding regions of the face (maxilla, eye, lips, nose, mandibula), the frequency of paroxysms can increase or the pain can present more continuously and oppressively. Hunt also reported a prosopalgic form of geniculate neuralgia, a more continuous pain, localized deeply in the hemifacies and sometimes irradiating to the ear.

When refractory to medical treatment (carbamazepine, phenytoin, lamotigrine or baclofen), surgery of the innervating nerves of the ear (the fifth, seventh, ninth and Xth cranial nerves and upper cervical dorsal roots) can be considered. Two main surgical techniques can be distinguished. Some surgeons have sectioned the intermediate nerve (Clark and Taylor, 1909), the chorda tympani (Rosen, 1953), the geniculate ganglion (Pulec, 1976), the glossopharyngeal nerve (Reichert, 1933), or performed a combination of these procedures. Other surgeons performed a neurovascular decompression of the forementioned nerves (Lillie, 1936).

We present a case report of geniculate neuralgia secondary to an occipital dysraphism.

Case report

This 35-year-old female complained of chronic geniculate neuralgia at the right side. The only relevant medical antecedent was the resection of an occipital subcutaneous lipoma in childhood. The last year this neuralgia had

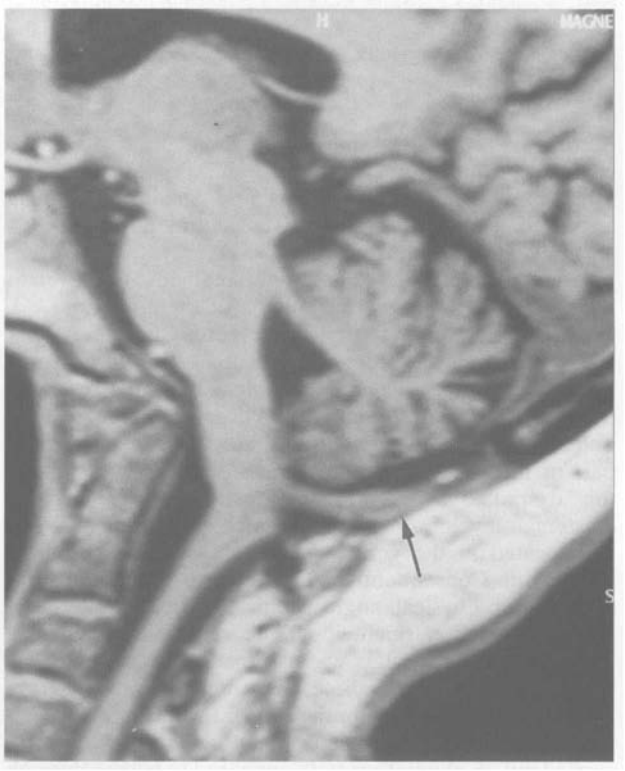

FIG. 1
Sagittal T1 weighted spin-echo image revealing brain stem tethering.

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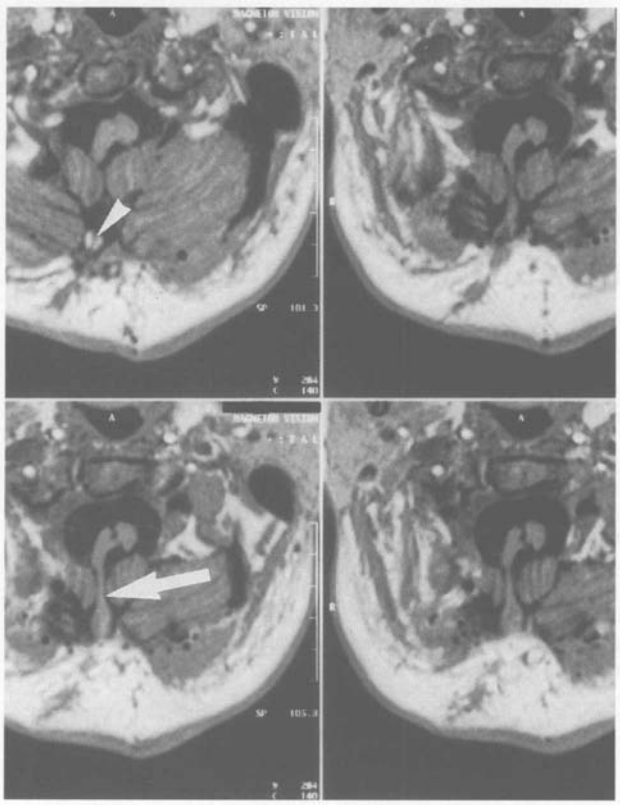

FIG. 2

Axial magnetization prepared rapid acquisition with gradient echo images. The dermal sinus tract can be seen extending from a cutaneous ostium towards the medulla oblongata, at the junction with the cervical spinal cord (arrow). Note the cervical diastematomyelia and the associated dermoid cyst (arrowhead).

become more frequent and intense and could be triggered by tactile stimulation of the right auditory canal. At the same time, a more continuous and less sharp otalgia appeared. Tricyclic antidepressives, neuroleptics, baclofen and phenytoin did not prove to be effective. Carbamazepine initially alleviated the pain but finally did not, even at high doses. MRI, organized by one of the authors (J.C.), revealed an occipital dysraphism (Figures 1 and 2). A stalk appeared to extend from the subcutis – through a small bone defect – to the medulla oblongata. This tethering mechanism seemed to cause a displacement of the medulla oblongata in a left posterolateral direction. The intradural part of the stalk contained a small dermoid cyst. Finally this dysraphic syndrome also presented as a cervical diastematomyelia. Otherwise a small cavernous angioma was located in the vermis and there was also a basilar impression. Consequently the patient was referred to our department. Physical, angiographical and electrophysiological examination (including blink reflex) were normal. The geniculate neuralgia was assumed to be provoked by traction on the intermediate nerve and/or cranial nerves IX–X, secondary to the brainstem distortion.

A posterior fossa trepanation and laminectomy C1 was carried out. A gliomatous stalk appeared to course through a small bone defect, intradurally below the vermis and finally it was attached to the medulla oblongata, inferior to the obex. A dermoid cyst was also identified and appeared to be fixed to the dura mater at the C1 level. The stalk was enveloped by arachnoid tissue, dissection was performed following this layer. We noticed a torsion of the floor of the fourth ventricle and the medulla to the left posterolateral, causing traction and torsion on the right cranial nerves IX, X and XI. No neurovascular conflict was seen. Finally, the stalk was resected, resulting in a reposition of the brain stem in a more anatomical position.

Anatomopathological examination revealed mixed gliomatous and fibrous tissue. Post-operatively there was a marked decrease of the geniculate neuralgia. Nevertheless, the first three post-operative months were characterized by prolonged vomiting. It was assumed that this was due to a reposition of the brain stem, resulting in a changed anatomical course of the lower cranial nerves. This complaint diminished spontaneously. After two years follow-up the patient estimated the geniculate neuralgia to be 20 per cent of the pre-operative intensity and she was able to stop the analgesic medication.

Discussion

Before considering the diagnosis of geniculate neuralgia, an extensive differential diagnosis should be considered. Acute otitis media and externa, adenoid cystic carcinoma of the external auditory canal, nasopharyngeal and laryngeal tumours, an elongated styloid process, temporomandibular joint disease, dentological pathology, thyroid tumours and tumours in the cerebellopontine cistern should be excluded. The aetiology of primary geniculate neuralgia is not known. Some authors advocate the hypothesis of a neurovascular conflict (Jannetta, 1975), supposing that the chronic compression by a vascular structure injures cranial nerves. Other authors contradict this theory, since neurovascular conflicts have been found during autopsies in asymptomatic patients and since sometimes radiologically neurovascular conflicts have been found on the contralateral side of the symptoms.

The treatment of this primary geniculate neuralgia is in the first instance a medical one. On failure of drugs such as certain anti-epileptics and baclofen, surgical therapy can be carried out. Only a few larger series have been published so far: Pulec (1976) sectioned the geniculate ganglion and/or the intermediate nerve via a middle fossa approach, obtaining relief in 13 of the 15 cases. Some patients underwent multiple procedures. Rupa *et al.* (1991) reported on 18 cases. Thirty-one operations were performed, most of them combining a neurovascular decompression with sectioning of the geniculate ganglion, the intermediate, ninth and/or 10th nerve. After a follow-up period of 3.3 years a 72 per cent success rate was obtained. Lovely and Jannetta (1997) finally published an experience with 14 patients. A retromastoid craniectomy was performed in order to decompress cranial nerves V, IX, X combined with sectioning of the intermediate nerve in most of the cases. A long-term follow-up was available in 10 patients and good results were found in 90 per cent of the patients. All three authors stress the complex sensory innervation of the ear, obliging the surgeon to inspect cranial nerves, V, VII_b, IX and X during the procedure.

In our patient, MRI revealed an occipital dysraphism as the cause of the geniculate neuralgia, leading to a particular surgical approach. A posterior fossa craniectomy confirmed an obvious tethering and distortion of the brain stem, and traction on the right cranial nerves IX, X and XI. After untethering, a reposition of the brain stem was noticed leading to some relaxation of the forementioned cranial nerves. After a follow-up period of two years, a marked decrease of the geniculate neuralgia is still experienced by our patient.

Previously, only three cases of brain stem/cerebellar tethering had been reported. The first case (Comey and Albright, 1994) concerned a midline occipital encephalocele, which had been transsected in the neonatal period.

Eight months later, an associated hydrocephalus necessitated a ventriculocardial drainage. The patient was doing well until the age of 24 when he developed drop attacks, sensory disturbance in the four limbs, incoordination in both hands, and positional headache, evoked by neck anteroflexion. MR and computed tomography (CT) scanning demonstrated tethering of the vermis. Cervical radiological examinations showed an associated Klippel Feil syndrome. An operative exploration confirmed the stalk, which was consequently excised, resulting in a resolution of the symptoms. Anatomopathological examination showed abnormal gliotic cerebellar tissue within fibrotic leptomeninges. Liu *et al.* (1995) reported a patient with a Dandy-Walker syndrome, treated by a cysto- and ventriculoperitoneal shunt. During the next four months multiple shunt revisions were carried out, finally the cyst drainage was abandoned. Nevertheless, at the age of four years a regrowth of the fossa posterior cyst required a replacement of the shunt. After a last revision of the shunt, the child developed – at the age of five years – progressive occipital headache and unilateral trochlear and abducens nerve dysfunction. In this case, a causal tethering of the lower brain stem was sectioned, the cyst was fenestrated and the cystoperitoneal shunt was removed. This untethering immediately improved the headache and cranial nerve deficits. Finally, Kernan *et al.* (1996) presented an 11-year-old boy with progressive gait ataxia, abasia, diminished proprioception in the four limbs, nystagmus, dysarthria, hypoglossal nerve dysfunction and urinary incontinence. On the first day of life, a Chiari type III malformation had been repaired and one month later a ventriculoperitoneal drainage had been implanted to treat hydrocephalus. MRI demonstrated tethering of the hindbrain at the cervicomedullary junction. Untethering resulted in improvement of the symptoms and signs, but this patient did not regain the previous neurological and functional level.

These case reports prove the importance of performing MRI before considering the primary repair of craniocervical meningoceles and encephaloceles. Whenever tethering is suspected, an intradural exploration is necessary and afterwards the dura mater should be reconstructed carefully, preventing secondary tethering.

Conclusion

Hindbrain tethering can be a rare cause of geniculate neuralgia. MR imaging proved to be an essential diagnostic procedure in the therapeutic planning and the surgical approach of this rare disease. In our case an untethering of the brain stem resulted in a marked decrease of the geniculate neuralgia during a follow-up period of two years.

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