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Unusual Painful Trigeminal Neuropathy Caused by Racemose Neurocysticercosis

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Dear Editor,

Space-occupying lesions in the cerebellopontine angle (CPA) and pre-pontine cistern can manifest as painful trigeminal neuropathy. Cases of meningiomas, schwannomas, arachnoid cysts, and tuberculomas causing trigeminal neuralgia have frequently been reported. In some series, up to 8.5% of cases of trigeminal neuralgia are related to tumors, especially those involving the CPA [1].

On the other hand, cysts of racemose neurocysticercosis are often located in the basal cisterns and CPA. In these cases, the symptoms are related to inflammatory reactions caused by the host–parasite interactions, and patients can develop headache, cysticercal meningitis, vasculitis, and hydrocephalus (due to mechanical obstruction of cerebrospinal fluid flow or arachnoiditis of the basal cisterns) [2]. Despite their proximity to cranial nerves, racemose cysts rarely produce symptoms related to compressive neuropathy [3].

We report the case of a patient with racemose neurocysticercosis who presented with painful trigeminal neuropathy that was alleviated after microsurgical removal of the cysts.

A 64-year-old man previously diagnosed with systemic hypertension was referred to our department with an 8-month history of facial pain on the right side. The patient described the pain as intense, lancinating, shock-like, and paroxysmal, with several daily attacks. There were no precipitating factors, and the pain was worsened by light touch to the affected side of the face and by mastication. The pain involved the second and third divisions of the right fifth cranial nerve. Physical examination revealed hyperesthesia and allodynia in the affected divisions of the trigeminal nerve.

The patient was previously managed with medical treatment, but did not experience any significant improvement of symptoms. Additionally, he could not tolerate the side effects of high doses of carbamazepine, which caused intense somnolence.

Magnetic resonance imaging revealed multiple cysts in the pre-pontine cistern and near the root entry zone of the right trigeminal nerve in the pons (Figure 1). Cerebrospinal fluid (CSF) analysis showed a protein level of 50 mg%, glucose of 55 mg%, and no leucocytes/mm [3]. Microbiological examination was negative. ELISA was positive for anticysticercus IgG.

Microsurgical 3removal of the cysts was proposed. A right retrosigmoid suboccipital craniotomy was performed. During cerebellar retraction for CSF drainage, a large cyst spontaneously exited the cistern and was removed. The remaining cysts were removed cautiously (Figure 2). At the end of surgery, no compression of the trigeminal nerve was observed, particularly at the root entry zone.

The patient had resolution of symptoms after surgery, and the medication was progressively ceased.

Neurocysticercosis is the most common parasitic disease of the central nervous system in the world. The disease is endemic in many developing countries, and migratory flows have resulted in new cases in the United States and in Europe [4]. The clinical presentation of neurocysticercosis is pleomorphic and depends on the number, stage of development, and location of the parasites in the central nervous system. Cysts found in the CSF compartments determine the racemose form of neurocysticercosis, which is characterized by a chronic inflammatory reaction that causes headache, meningitis, hydrocephalus, and intracranial hypertension [2].

Despite the proximity of cysts to the cranial nerves in the posterior fossa, symptomatic neuropathies are uncommon. An extensive literature review identified only five previous cases. In 1963, Tenuto et al. [5] reported two cases of trigeminal neuralgia due to mechanical compression caused by racemose cysts, but the cysts were located in Meckel's cave. In 1995, Revuelta et al. [3] described the case of a patient with trigeminal neuralgia on the right side and a cerebellopontine cyst on the left side. The authors argued that the mass effect from the cyst distorted the brainstem to the right, compressing the nerve against an arterial loop at the root entry zone. The pain disappeared soon after surgical resection of the cyst. In 2003, the same authors reported the case of a patient with a racemose cyst in the CPA and concomitant vascular compression of the fifth nerve at its entry zone. In this case, microsurgery permitted excision of the cyst and arterial decompression of the nerve, with the resolution of symptoms [6]. In 2000, Aguiar et al. [7] described the case of a patient with bilateral trigeminal neuropathy, that was more intense on the right side. Microsurgical removal of a racemose cyst in the right CPA provided relief of the bilateral symptoms.

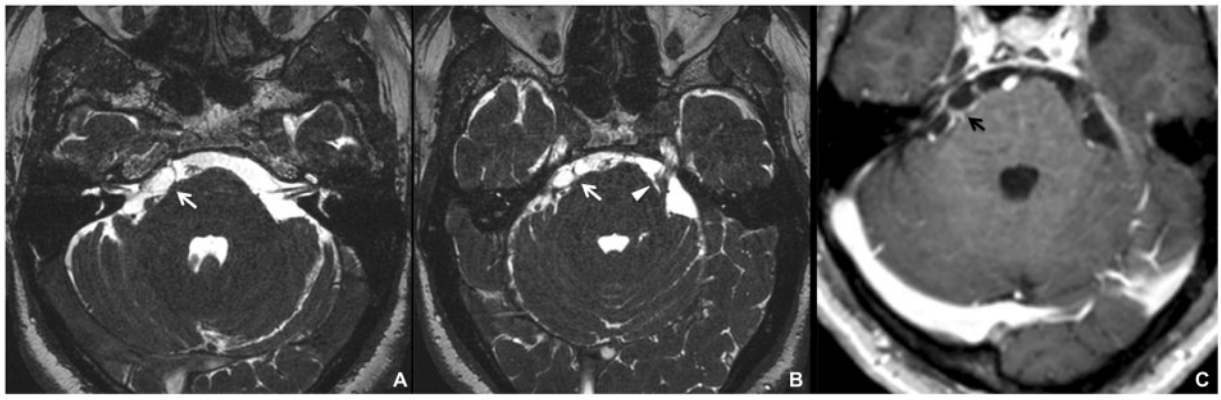

Figure 1 Magnetic resonance imaging (fast imaging employing steady-state acquisition) showing multiple cysts in the posterior fossa cisterns (arrows), mainly in the cerebellopontine angle (A), which caused compression of the trigeminal nerve on the right. Compare with normal left fifth nerve (arrowhead, B). The cysts were enhanced on T1 axial image (black arrow, C).

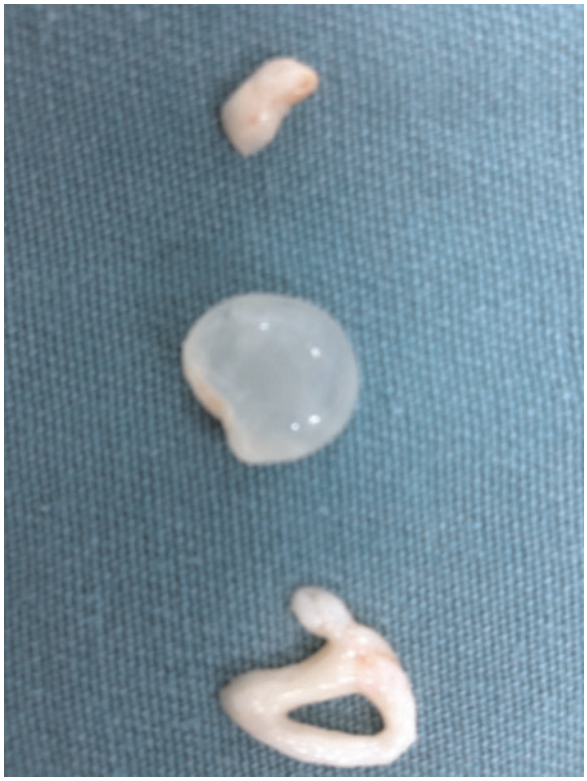

Figure 2 Cysts removed from the cisterns of the posterior fossa. Histopathological analysis confirmed that the lesions were cysticerci.

Despite general similarity to the five previously reported cases, the present case is unique, as the patient presented the cysts on the same side as the pain and no vascular compression was observed. We believe that the cysts produced direct compression of the nerve, which can occur with other tumoral lesions in the region even in the absence of concomitant vascular compression. Therefore, the patient's pain pattern met the criteria for painful trigeminal neuropathy attributed to space-occupying lesions described by the International Headache Society [8].

Neuroimaging is useful in the diagnostic workup of trigeminal neuropathy [9] It is important to rule out other causes of pain, such as space-occupying lesions, which require a specific approach rather than just empiric medications. In our case, the recognition of cysts within the CPA guided the adequate treatment.

One may argue that medical treatment with anthelmintics should be the first option for the management of cysts within the CPA, as long as the drug is able to destroy the parasites and remove the compression factor from the nerve. However, in fact, the use of anthelmintics for the management of extraparenchymal neurocysticercosis is controversial [10], as the death of the parasites exacerbates local inflammatory responses. We therefore believe that the local damage to the nerve may have worsened the pain in our case. Actually, consensus on the management of neurocysticercosis points to the removal of cysts when possible, followed by antiparasitic treatment [11]. Degenerating cysts may be adhered to the cranial nerves and vessels and, in these cases, surgical removal is hazardous. On the other hand, we believe that the microsurgical removal of active cysts involving the CPA is safe, minimizes the risk of nerve damage, and provides immediate relief of symptoms in the case of symptomatic neuropathy.

To our knowledge, this is the sixth reported case of painful trigeminal neuropathy attributed to racemose neurocysticercosis. However, in contrast to

previous reports, the typical clinical presentation of trigeminal neuralgia was directly related to the side and place of compression of the fifth nerve in the absence of concomitant vascular conflict. We highlight the need for diagnostic imaging as part of the workup for trigeminal neuropathy and the effectiveness of microsurgery for the removal of cysts and pain relief.

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