Trigeminal Neuralgia Caused by a Choroid Plexus Papilloma of the Cerebellopontine Angle: Case Report and Review of the Literature

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This report describes a 59-year old woman with a rare choroid plexus papilloma of the cerebellopontine angle presenting with trigeminal neuralgia. The patient was admitted complaining of a 12-year history of paroxysmal lancinating pain throughout the right side of her face. Treatment with carbamazepine, Chinese medicine and a peripheral neurectomy had not relieved the pain. At operation, a 0.5 × 0.5 × 0.5 cm neoplasm was found in the cerebellopontine angle, which was firmly adherent to the roots of the seventh and eighth cranial nerves and the brainstem. There was no apparent tumour bulk or vascular compression around the trigeminal nerve root entry zone. Subtotal tumour excision and selective partial rhizotomy were performed. The patient's facial pain gradually resolved. Involvement of the trigeminal nucleus in the brainstem by the cerebellopontine angle tumour is suggested as the possible cause for trigeminal neuralgia in this case.

KEY WORDS: CHOROID PLEXUS PAPILLOMA; TRIGEMINAL NEURALGIA; CEREBELLOPONTINE ANGLE

Introduction
Trigeminal neuralgia generally results from mechanical compression of the trigeminal nerve, causing irritation and change in neural function that ultimately leads to hyperactivity of the trigeminal nerve nucleus. The cause of trigeminal neuralgia is not always certain. Typically, the pain is associated with dislocated blood vessels pressing onto the nerve.1 Cerebellopontine angle tumours that cause compression or traction of the trigeminal nerve are uncommon,2–4 but are a recognized cause of secondary trigeminal neuralgia. In some cases, trigeminal neuralgia may result from other disorders, such as multiple sclerosis or infection.5 This report describes a case of cerebellopontine angle choroid plexus papilloma in a 59-year old woman presenting with trigeminal neuralgia.

Case report
A 59-year old woman was admitted to the Department of Neurosurgery, Qilu Hospital of Shandong University, Jinan, China, complaining of a 12-year history of paroxysmal lancinating pain throughout the right side of her face. She had been treated with carbamazepine and Chinese medicine, and had undergone a peripheral...
neurectomy of the affected trigeminal branch 6 years previously. This had not, however, relieved the pain.

Neurological examination demonstrated very mildly diminished sensation in the trigeminal nerve V3 distribution on the right side. The remainder of the neurological examination was normal. The pre-operative brain magnetic resonance image (including a scan after gadolinium diethylenetriaminopenta-acetic acid injection) showed no abnormalities.

A right retromastoid approach to the trigeminal nerve root was performed with the patient in the semi-prone park bench position. With the cerebellum retracted caudal and backward, the sensory and motor branches of the trigeminal nerve were located after sharply dissecting the arachnoid membrane. Unexpectedly, a soft $0.5 \times 0.5 \times 0.5$ cm reddish neoplasm was found in the cerebellopontine angle, which was firmly adherent to the roots of the seventh and eighth cranial nerves and the brainstem. There was, however, no apparent tumour bulk or vascular compression around the trigeminal nerve root entry zone. The mass was subtotally resected piece by piece because of its firm attachment to the brainstem. Rhizotomy of the inferior one-third of the sensory portion of the trigeminal nerve was performed for relief of the facial pain.

The patient recovered without severe complications or further post-operative neurological deficits. The patient’s facial pain resolved gradually after the operation. At the 2-year follow-up examination, the patient described no pain. Histological examination of the resected mass showed it was a choroid plexus papilloma (Fig. 1).

**FIGURE 1**: Choroid plexus papilloma showing the typical papillary pattern comprising a single layer of columnar cells (haematoxylin and eosin staining)
Discussion

Trigeminal neuralgia is caused by cerebellopontine angle tumours in only 5 – 10% of cases. Such tumours are most frequently epidermoid tumours, acoustic neurinomas or meningiomas; occasionally trigeminal neuralgia has been associated with other posterior fossa tumours such as lipomas, metastases, lymphomas or glioblastomas.2,3,6 Choroid plexus papillomas are rare neoplasms of neuroectodermal origin occurring in adults. About 35 cases of cerebellopontine angle choroid plexus papilloma have been described in the literature.7 – 9 All of them presented with signs of increased intracranial pressure and cranial nerve deficits, such as headache, vomiting, dizziness, unstable gait, hearing loss or hemifacial spasm. An extensive review of the major series and case studies reporting cerebellopontine angle choroid plexus tumours found no report of a choroid plexus tumour causing trigeminal neuralgia. To the best of our knowledge, the case presented here is the first case of trigeminal neuralgia caused by a cerebellopontine angle choroid plexus papilloma.

The exact mechanism underlying trigeminal neuralgia in cerebellopontine angle tumours remains unclear; many different hypotheses have been proposed. In epidermoid tumour, acoustic neurinoma or meningioma of the cerebellopontine angle, the most common cause of trigeminal neuralgia has been suggested to be direct compression of the nerve by the tumour or an inflammatory reaction elicited by the tumour.10 Infiltration of the trigeminal nerve has been proposed as another cause of neuralgia in lipoma and lymphoma of the cerebellopontine angle.5,11 Other suggested mechanisms include stretching of the trigeminal nerve, neurovascular conflict and local ischaemia of the brainstem subsequent to compression by a large cerebellopontine angle tumour.12 In the present case, the choroid plexus papilloma did not directly compress the trigeminal nerve and there was no evidence of neurovascular conflict at the root entry zone of the trigeminal nerve at operation. Because of the tiny bulk and soft texture, the tumour could not have led to stretching of the trigeminal nerve by distorting the brainstem. Thus, the present case does not fit well with any of the hypotheses mentioned above.

In recent years, the ‘central’ hypothesis that trigeminal neuralgia is caused by disease in the ‘central’ nervous system has been proposed. Rosetti et al.13 suggested that involvement of the nucleus of the spinal trigeminal tract and the principal sensory nucleus of the trigeminal nerve with segmental demyelination of the trigeminal nerve fibres may be responsible for the development of trigeminal neuralgia. Caranci et al.14 reported that chronic extrinsic compression on the trigeminal pain pathway by herniated cerebellar tonsils was the neuropathological basis of facial pain in patients with Arnold–Chiari type I malformation. A distinctive feature of the tumour in the present case was the firm attachment to the brainstem, where the nucleus of the spinal trigeminal tract and the principal sensory nucleus of the trigeminal nerve are located. We propose that compression of the trigeminal nucleus in the brainstem by the cerebellopontine angle choroid plexus papilloma played a central role in the pathogenesis of trigeminal neuralgia in this patient by increasing excitation and decreasing inhibition in the trigeminal nuclear complex.

In conclusion, this case report serves as a reminder that a tiny cerebellopontine angle tumour may be responsible for trigeminal neuralgia, even if the magnetic resonance
image shows no abnormality. In such cases, neurosurgical decompression is essential for pain relief. In order to relieve neuralgia, partial rhizotomy should be performed if the tumour cannot be totally resected because of its firm attachment to the brainstem.

Conflicts of interest
The authors had no conflicts of interest to declare in relation to this article.

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