A case of trigeminal neuralgia caused by intracranial epidermoid tumor

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Abstract Background: Cerebellopontine angle epidermoid is a rare congenital tumor. This is a slow growing benign tumor with late presentation in adulthood. There is a possibility of missed diagnosis. Case Presentation: A 56 year-old woman had intermittent left cheek pain for 10 years. The patient was diagnosed with typical trigeminal neuralgia (TGN) at a general hospital. She was treated with a satellite ganglion block and oral medication. As a typical trigeminal neuralgia, the nature of pain was atypical. During the first examination, sensory disturbances in the second branch of the trigeminal nerve in the left side and delayed light reflexes of the left pupil were detected. Cranial magnetic resonance imaging (MRI) was performed. The result of the MRI showed a cerebellopontine angle epidermoid tumor. TGN resolved completely after near complete tumor resection. Upon removal of the tumor, the patient required no analgesics. Conclusion: We reported a case of TGN due to epidermoid. The diagnosis was made after a thorough neurological examination. Regular MRI is also necessary. This is because symptoms are exhibited even if the tumor is small. To ensure correct diagnosis and treatment, careful case management in the pain clinic is necessary.

Keywords: epidermoid, painful trigeminal neuropathy, space occupying lesion, trigeminal neuralgia, intracranial tumor

1 Introduction

Trigeminal neuralgia (TGN) is a syndrome consisting of unilateral short lancinating pain in the distribution of one or more branches of trigeminal nerve. Pathogenesis of TGN includes vascular compression of trigeminal nerve root, intracranial tumors, intracranial infection and multiple sclerosis plaques in the medulla spinalis. The 5th nerve compression of any intracranial tumor may cause TGN. Epidermoid tumor located at cerebellopontine angle (CPA) is rare and may cause TGN. Intracranial epidermoid tumor is slow-growing. Even small epidermoid also cause symptoms because symptoms were induced by not only nerve compression but also aseptic meningitis caused by the leakage of the cholesterol crystal from tumor. In patients with long term TGN treatment, therefore regular neurological examinations and appropriate neuroimaging study are essential to avoid overlooking a small tumor in the CPA cistern.

In preparing this report, written consent was obtained from the patient herself.
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II Case Report

A 56-year-old woman presented with the chief complaint of left cheek pain of 10 year’s duration. The pain was sharp, intermittent on the left cheek and provoked by teeth brushing and making up. She was diagnosed with trigeminal neuralgia at a general hospital when she had had first attack. She had been treated with satellite ganglion block and medication there.

That symptoms appeared and disappeared every half to one year. Since she had a symptom for the first time in a year, she visited our clinic. Sharp pain spread from the maxilla to the forehead with non-noxious stimulation to the upper alveolus and left upper lip. That pain lasted around half an hour once the pain attacked.

Evaluation of her past history revealed systemic lupus erythematosus and nephrosis syndrome. She was taking pregabalin 300 mg, prednisolone 5 mg, tablet formulated with olmesartan/azelnidipine, atorvastatin 10 mg, omeprazole 20 mg, clopidogrel 75 mg. She came out in a rash with oral carbamazepine administration. On neurologic examination, hypoesthesia in 2nd branch of trigeminal nerve area in left side and delayed in light reflex of the left pupil were recognized. Including diplopia, other neurologic abnormal findings were not found.

The pain was too strong to make speech and have meals. She was undertaken infraorbital nerve block with 0.5% levobupivacaine 3 ml. After that nerve block, the pain was relieved for 6 h. Because she had neurological abnormality, she underwent cranial magnetic resonance imaging (MRI), which demonstrated an intracranial tumor anterior to the brainstem. The tumor was hypointense on T1-weighted images and hyperintense on T2-weighted images (Figure 1a). The tumor was hyperintense on diffusion-weighted images (Figure 1b). On MRI cisternography (constructive interference in steady state: CISS), the tumor was located along the course of troclear nerve, trigeminal nerve, facial and vestibulocochlear nerves on the left. The tumor distorted trigeminal nerve laterally (Figure 2).

The patient consulted neurosurgery and underwent operation with preoperative diagnosis of epidermoid tumor. As for the facial nerve and the inner ear nerve, they were asymptomatic and therefore sharp separated from the tumor. Trigeminal nerve and the tumor were strongly adhered, so the surgery was ended leaving a part. The tumor was almost completely resected. On removing the tumor, she required no analgesic.

III Discussion

Trigeminal neuralgia is common disease, while the incidence of trigeminal neuralgia due to intracranial tumor is less than 0.8%. The Brain Registry of Japan found that the incidence of intracranial epidermoids accounted for

Figure 1  MRI shows intracranial tumor anterior to the brainstem in left side
a: Left: Tumor with hypointense is recognized on T1-weighted image. Right: Tumor with hyperintense is recognized on T2-weighted image.
b: Tumor with hyperintense is recognized on diffusion-weighted image.
1.4% of all primary brain tumors. In that study, 252 epidermoid cases (6.3%) were registered as CPA tumors, as well as 3,632 cases of neuromas (90.5%) and 639 cases of meningioma (15.9%). The incidence of TGN in patients with CPA epidermoids has been reported to vary from 0 to 76.9%. According to Kobata et al., 28 (93.3%) of 30 patients presented with TGN associated with epidermoids in Japan. Epidermoid, the third most common tumor, is taken into account at the incidence of CPA tumors. The inflammation because of the leakage from tumor causes TGN, small tumor that do not appear in MRI can cause symptoms. Regular neurological examination and appropriate neuroimaging tests are essential, as it may cause neurological findings while the treatment course is prolonged.

Epidermoid arises during 3rd to 5th week of embryogenesis due to incomplete separation of neuroectoderm from surface ectoderm. Although this tumor is commonly presented in CPA, some reports were presented in pontine, ventricles and parasellar region. Epidermoids are covered with stratified squamous epithelium and contain keratin, cell debris and cholesterol. Intracranial epidermoids usually spread and adhere to the critical neurovascular structures along the cistern. Clinical symptoms include TGN, headache, dizziness, progressive hemiparesis, unstable gait and hemifacial spasm. Lagares et al. revealed that compression injury by epidermoid to the trigeminal nerve root leading to demyelination is a major determinant in the pathogenesis of TGN according to CPA epidermoid. Additionally, substances that leak from the tumor stimulate nerve roots and cause symptoms. In this case, since only the trigeminal nerve had adhered to the tumor, it is expected that trigeminal nerve showed symptoms. Trigeminal nerve involves not only motion fibers and sensory fibers to the face but also pupilloconstricting fibers.

Because epidermoid tumor grows extremely slowly, the average age of surgically treated patients was in their forties. Age at onset of TGN and operation of epidermoid TGN are earlier than vascular compression TGN. The patients’ average ages at the onset and at the time of surgery were 37.8 and 54.1 years, 49.3 and 60.4 years, respectively. In young patients with TGN, therefore, the neuroimaging study must be performed carefully to avoid overlooking a small tumor in the CPA cistern.

The opportunity to be diagnosed with CISS for TGN...
has been increased. MRI T1- and T2-weighted images showed epidermoids as hypointense and hyperintense, respectively, compared with cerebrospinal fluid and brain tissue. Diffusion-weighted images provided more useful for diagnostic evaluation.

Surgery is the first choice for treatment of CPA epidermoid. Thirty percent of patients with subtotal removal experienced symptomatic recurrences after 8.1 years after surgery, whereas all patients with total removal were still asymptomatic. The recurrence-free survival rate was 95% at 13 years for patients with total removal compared with 65% for patients with subtotal removal. In this case, we can obtain temporal pain relief after the nerve block. Nerve block may obtain pain relief at the time of recurrence of epidermoid TGN in older ages and high risk cases.

In this case, epidermoid could be diagnosed with careful neurological examination at the first visit and appropriate image examination. If the course of treatment is long-term, it is necessary to take neurological findings and take appropriate diagnostic images into account. Especially, when it is against the diagnostic criteria, we need to pay sufficient attention. MRI examination including CISS and diffusion-weighted image is actively performed when suspecting CPA epidermoid tumor.

References