Epidermoid Cyst Presenting as Isolated Trigeminal Neuralgia - Two Case Reports

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Abstract-

Purpose: Symptomatic TN accounts for up to 15% of all TN. Though there are many established "red flag" signs, it is still sometimes difficult to sift symptomatic from classic TN. We herein report two cases of isolated TN with normal neurologic examinations and then tissue proved as epidermoid cyst.

Case Report: **Case 1:** A 17-year-old girl presented with paroxysmal intense pain mixed dull background pain at right mandibular region for one month. The blink reflex demonstrated brainstem lesion and brain magnetic resonance imaging (MRI) revealed a huge lobulated tumor in right cerebellopontine angle (CPA) with obvious brainstem compression. Her right facial pain was nearly completely disappeared postoperatively. **Case 2:** The 48-year-old woman had chronic paroxysmal electric-like and burning pain in left V3 region for more than 5 years. Because of refractory pain, brain MRI was arranged and showed a non-enhancing cystic lesion at left CPA. Post operative complications occurred as left multiple lower cranial nerve palsies and Horner syndrome, and truncal ataxia. Her facial pain was completely free after 1 month follow up.

Conclusion: In the first patient, teenage onset, abnormal trigeminal reflex, and early developing background pain struck us directly to symptomatic TN. In the second case, we suspected symptomatic TN with uncertainty before image study. TN could be the isolated initial symptom of CPA epidermoid cysts. In consideration about pretty high prevalence of symptomatic TN, physicians should be more alert and straightforward arrange neuroimage when facing TN patients with atypical presentation.

Key Words: epidermoid cyst, trigeminal neuralgia, cerebellopontine angle

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INTRODUCTION

Trigeminal neuralgia (TN) is the most common craniofacial pain syndrome, and is characterized by unilateral stereotyped attacks of intense and short-lasting pain that affect one or more divisions of the trigeminal nerve. It occurs more in females, with an annual incidence of 4-5/100,000[(1)], and is commonly precipitated by touching trigger areas while chewing, eating, or brushing teeth. 70% of patients are older than 60 years

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at onset[2].

TN can be classified as classic or symptomatic. The pathogenesis of classic TN is idiopathic or attributable to vascular compression of the trigeminal nerve roots[3], whereas symptomatic TN is caused by a structural lesion[1]. These lesions include tumors, multiple sclerosis, and pontine infarction. In clinical practice, differentiating symptomatic from classic TN is important but sometimes difficult. We herein report two cases of isolated TN with the final diagnosis of symptomatic TN.

CASE REPORT

Case 1

A 17-year-old female presented with paroxysmal intense pain and paresthesia at the right mandibular region for one month. The pain involved the right V3 division only, and was aggravated by speaking and swallowing. Each episode lasted for seconds to one minute, and the attack was stereotyped. Though the clinical course was only one month, the dull background pain developed between the more severe attacks.

After evaluation by a dentist, she was referred to our neurology clinic where the neurological examination findings were all unremarkable. Due to the teenage-onset and persisted background pain, symptomatic TN was highly suspected. The blink reflex stimulated on right supraorbital nerve showed prolonged latency of R1 and normal R2 on both sides, while stimulated on the contralateral nerve showed normal R1 on the left side and normal R2 on both sides. This result indicated right pontine lesion (Fig. 1). Brain magnetic resonance imaging (MRI) revealed a large, lobulated, extra-axial tumor at the right-side cerebellopontine angle (CPA) with brainstem compression (Fig. 2A). She underwent a right retromastoid craniectomy with total removal of the tumor. During operation, the neuro-surgeon found that the tumor compressed the root entry zone of right trigeminal nerve, right facial and vestibulocochlear nerve. The final histopathology showed that the tumor contained mainly keratin material without epithelial lining, which is the hallmark of an epidermoid cyst (Fig. 2B). Two weeks after the operation, cerebrospinal fluid from the craniectomy wound had accumulated subcutaneously and the patient suffered from episodes of fever. This improved after local aspiration and temporary use of empiric antibiotics, and no new neurological deficits occurred. She remained completely free of facial pain and the cyst did not recur at the 1- year follow-up.

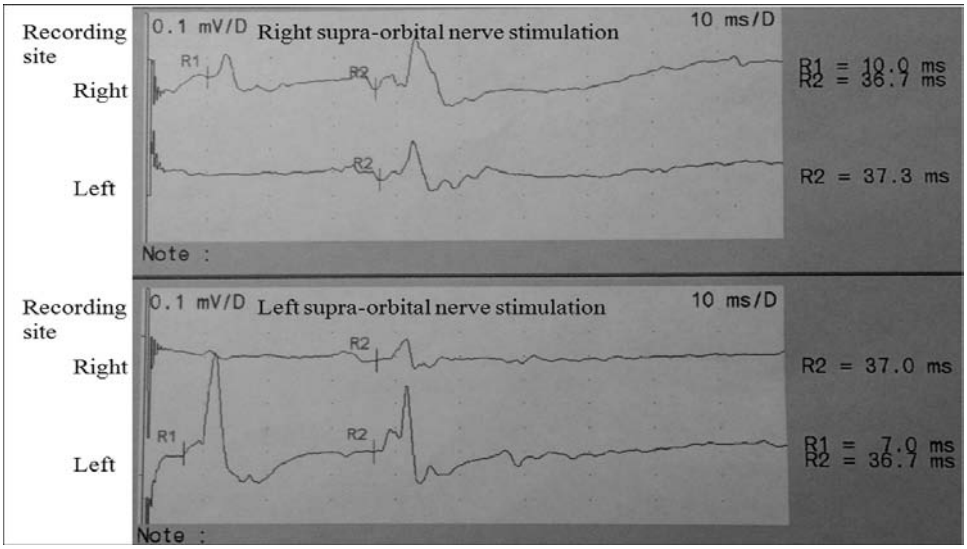

Figure 1. Blink reflex showing delayed R1 (10.0 ms) on right side and normal R2 (36.7 ms, right; and 37.3 ms, left) on both sides while stimulation on right supraorbital nerve; normal R1 (7.0ms) on left side and normal R2 (37.0 ms, right; and 36.7 ms, left) on both sides while stimulation on left supraorbital nerve.

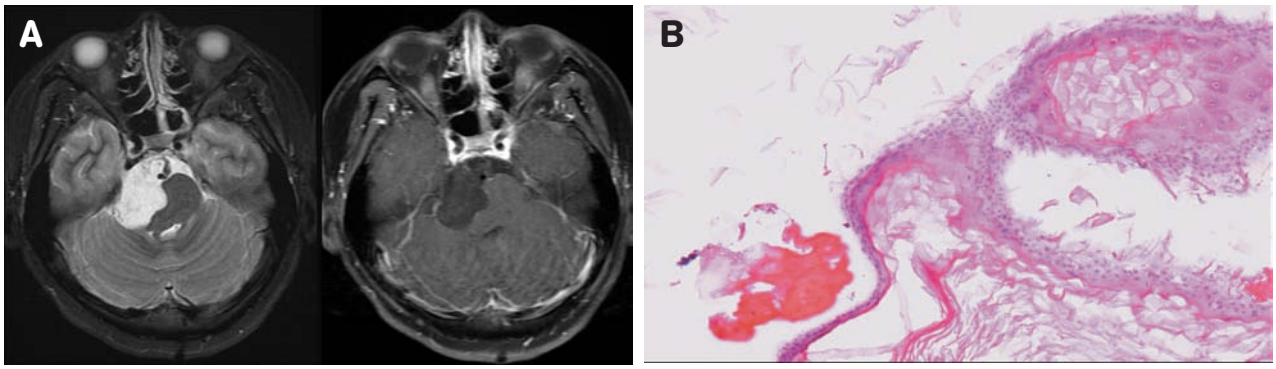

Figure 2. (A) Brain MRI revealed a large lobulated extra-axial (around 2.5 × 3.5 cm) tumor at the right cerebellopontine angle (CPA), with invagination into the medial aspect of the upper brainstem. The tumor engulfed the basilar artery and trigeminal nerve, and showed hyperintensity in T2-weighted imaging (A, left) and hypointensity in T1-weighted imaging, with no signal change after gadolinium enhancement (A, right). (B) The histopathology of the epidermoid cyst from case 1 (hematoxylin-eosin stain, original magnification × 100)

Case 2

A 48-year-old female had chronic intermittent stereotyped sharp, electric-like, burning pain extending from the left marginal tongue to the ipsilateral mouth angle, lower ear, and cheek for more than 5 years. Initially it only lasted for seconds, however she gradually experienced persistent background dull pain which was provoked by either eating or brushing her teeth, and only partially relieved by local pressing and medications from an otolaryngologic clinic. The pain was poorly controlled with carbamazepine 600 mg per day for 5 years. She then visited a neurologist due to refractory facial pain, however the neurological examinations were unremarkable and the blink reflex test showed normal results. Brain MRI with gadolinium enhancement demonstrated a non-enhancing cystic lesion at the left CPA and compression of the left brainstem (Fig. 3).

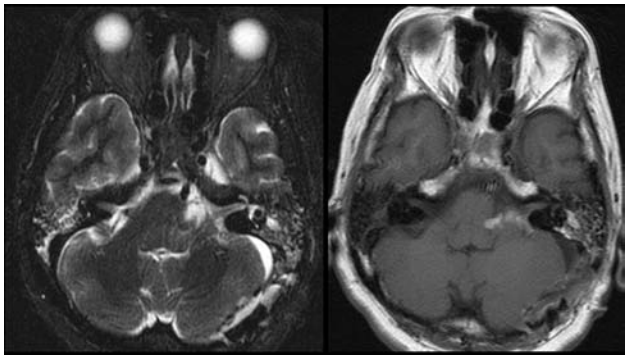

Figure 3. Brain MRI with gadolinium enhancement demonstrated a non-enhancing cystic (around 0.8 × 1.0 cm) lesion with diffusion restriction at the entry zone of trigeminal nerve and compression of the left brainstem, left V, VI, VII, VIII cranial nerves.

The patient hesitated to undergo surgery until 6 months later, when she experienced more severe pain and more drowsiness after shifting from carbamazepine to gabapentine 600 mg per day. Left retromastoid craniectomy with tumor excision was performed and histopathology revealed keratin-filled cysts lined with laminated keratinaceous material without adnexal structures, consistent with the final diagnosis of an epidermoid cyst. Postoperative complications occurred in the form of left multiple lower cranial nerve palsies, left Horner's syndrome, and truncal ataxia. Her neurological deficits improved slightly after hyperbaric therapy and rehabilitation programs. Although there were neurological sequelae, she was completely free of facial pain after 1 month of follow-up.

DISCUSSION

Both of the cases in this study had an epidermoid cyst presenting with isolated TN as the initial symptom. In the first patient, teenage-onset, abnormal trigeminal reflexes, and the early development of background pain prompted the diagnosis of symptomatic TN. Because

refractory facial pain persisted in the second patient, symptomatic TN was considered in clinical practice. There are many "red-flag" signs that can alert physicians to the possibility of symptomatic TN. Among them, trigeminal sensory deficits and bilateral involvement are the most solid and occur only in symptomatic TN[1]. Abnormal trigeminal reflexes have a high sensitivity (94%) and specificity (87%) for an increased possibility of symptomatic TN[1]. In the blink reflex of the first patient, stimulation on right supraorbital nerve showing delayed latency of right R1 and normal latency of bilateral R2 may indicate ipsilateral lesions involved chief trigeminal sensory nucleus in right pons blocked R1 pathway, but spared dorsal trigeminal tract that preserved bilateral R2 responses[4,5]. In second patient, maybe smaller lesion with only partial trigeminal sensory nucleus compression allowed some electric current passed and resulted in normal blink reflex study.

Though young-onset TN, as in our first patient, is traditionally regarded as red-flag sign, it is sometimes difficult to be judged in clinical practice since considerable overlap in age range between classic and symptomatic TN, particularly if the patients are in the fourth decade of life[1]. Patients with epidermoid-related TN have a younger age at onset and longer duration of symptoms than those with vascular compression-related (classic TN)[6,7]. This can be explained by the congenital origin and the extremely slow growth rate of epidermoid tumors. By evidence-based review, refractory pain is not an absolute "red-flag sign"[1]. However, on the other viewpoint, TN is the most common craniofacial pain syndrome, and routine brain image screening can identify up to 15% of structural lesions[1]. In consideration of such high prevalence of symptomatic TN, physicians should be alert and promptly arrange neuroimaging for TN patients with any atypical presentation, such as our second patient.

In two previous case series of 24 and 30 CPA epidermoid cysts, the mean age at diagnosis was 37.8-38.8 years, and the mean duration from onset of symptoms to diagnosis was 3.1-11.5 years[6,7]. According to a meta-analysis of 263 cases of CPA epidermoid tumors, the most common cranial nerve symptoms were eighth cranial nerve dysfunction (37.6%), followed by TN (29.7%), facial palsy (19.4%), and headache (17.9%) [7]. Other unique manifestations include glossopharyngeal neuralgia [8], mental status change, seizures[6], and even short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms[9].

Epidermoid cysts originate from disarranged ectodermal cells during neural tube closure in the embryonic stage [10]. Intracranial epidermoids can grow slowly and then compress neurovascular tissue with only minimal clinical symptoms. The most common locations of intracranial epidermoids are the CPA (40-50%)[7], the fourth ventricle, and suprasellar regions[11]. Intracranial tumors rarely cause TN (< 0.8%)[11]. Tumors resulting in TN are usually located in the posterior fossa, such as acoustic neuromas and epidermoid cysts, and some in the central fossa, such as meningiomas, schwannomas, and pituitary adenomas[11]. Among them, epidermoid cysts constitute only 1.0-1.4% of cases[7,11]. The incidence of CPA epidermoid cysts in patients with TN has been reported to range from 0.03 to 5.5%[7].

Both of our patients underwent total removal of the tumors by retromastoid craniectomy, which is the golden standard treatment for CPA epidermoid cysts, although subtotal removal is also justified if the tumor adheres densely to vital neurovascular structures[6]. Since epidermoids are not sensitive to radiation or chemotherapy[6], the adjuvant radiotherapy may be applied only for recurrent epidermoid cyst or epidermoid cyst with malignant transformation[12,13]. In terms of postoperative complications, the first patient suffered from cerebrospinal fluid leakage and subsequently recovered completely, while the second patient had permanent neurological damage. Both patients achieved remission from TN after surgery. According to a previous study[6], new neurological deficits after surgery occurred in 87.5% of patients (21/24), however most of the deficits (17/24) were transient and the patients achieved complete recovery later. Cerebrospinal fluid leakage and aseptic meningitis are well-known postoperative complications, with two studies reporting 25% of patients (6/24) in 6.2 years and 6.7% (2/30) in 11.5 years of follow-up having recurrent tumors[6,7]. Both studies reported that most patients had

good neurological outcomes after undergoing surgery. Although our first patient had a larger cyst, she had a better postoperative neurological outcome than the second patient. This may suggest that the duration from onset to diagnosis and the position of the tumor are more important for the prognosis than tumor size.

Although there are many established "red-flag" signs, it can still be difficult to differentiate symptomatic from classic TN. Early developing and persistent background pain should be considered a sign of symptomatic TN. Refractory TN with a relatively young onset warrants imaging studies. TN may be an isolated and initial symptom of CPA epidermoid cysts. Since total removal surgery is standard treatment for epidermoids and most patients can achieve a good neurological outcome, patients should undergo an operation as soon as possible.

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