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Isolated facial nerve palsy of peripheral type caused by an intrinsic brain stem tumor

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Introduction

Facial palsy of the peripheral type is generally seen in the pain clinic and is often treated with a stellate ganglion block. The most common cause of peripheral facial nerve palsy is Bell's palsy, although its etiology remains controversial. The diagnosis of Bell's palsy is usually made by exclusion of other conditions such as herpes zoster oticus (Ramsay Hunt syndrome), trauma (including skull base fracture and surgery), otitis media, and neoplasm [1].

Isolated peripheral facial nerve palsy of neoplastic origin is uncommon. We herein describe a case of peripheral facial nerve palsy which was initially diagnosed as Bell's palsy but was later found to be caused by an intrinsic brain stem tumor.

Case report

A 9-year-old boy presented to the Pediatric Department of our University Hospital in August 1990 with left facial weakness. His mother noticed the hyperemic conjunctiva and lacrimation of his left eye at the end of June. Consultation with the ophthalmologist revealed no abnormality in his left eye and the hyperemia improved with conservative therapy. In July, facial asymmetry became obvious. He was diagnosed as having Bell's palsy by a pediatrician in August and was referred to our pain clinic.

Upon examination, the patient had a left facial nerve palsy of the peripheral type (score of the facial paresis was 24/40), however, no other neurological deficits were seen. An audiogram failed to reveal a hearing abnormality.

Although repeated stellate ganglion block was given, his facial palsy progressed slowly over a 2-month period, suggesting an etiology other than Bell's palsy.

Magnetic resonance imaging (MRI) in September demonstrated a tumor in the left pons and brachium pontis extending into the left cerebellopontine angle (Fig. 1). The lesion was seen as a hypointense and hyperintense area on T1- and T2-weighted images, respectively. He was admitted to the Neurosurgical Department on September 29. The positive neurological findings on admission were Bruns' nystagmus, absence of left corneal reflex, decreased gag reflex, and mild trunkal ataxia, in addition to left facial nerve palsy. He underwent a wide suboccipital decompressive craniectomy, and biopsy of the tumor indicated low-grade glioma. In spite of postoperative radiation (60 Gy) and chemotherapy including Ranimustine and tumor necrosis factor, he died due to tumor progression 17 months from the time of his initial symptom.

Discussion

Eighty percent of peripheral facial nerve palsy cases represent idiopathic or Bell's palsy, of which approximately 20% can be demonstrated to have a specific etiology [2]. Peripheral facial nerve palsy with neoplastic origin is uncommon, and is estimated to be the cause in approximately 5% of all cases [3].

The diagnosis of Bell's palsy is unjustified unless an accurate history is taken along with a careful examination of the ear and central nervous system (CNS). The differential diagnosis of neoplastic facial palsy is vast

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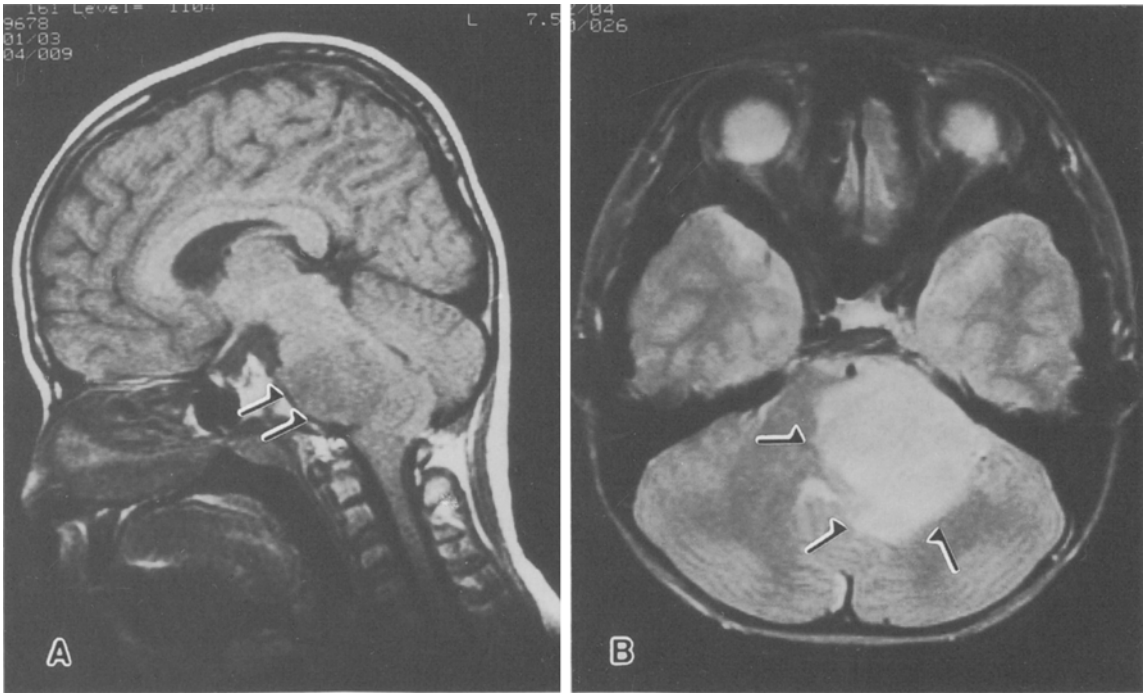

Fig. 1. **A** Pons is swollen with a hypointense lesion (*arrows*) on sagittal view of T1-weighted imaging. **B** Axial view of T2-weighted imaging demonstrates hyperintensity tumor (*arrows*) in the left pons and brachium pontis extending into left cerebellopontine angle

and depends upon a very complete neurological investigation by neurologists and neuro-otologists. In our case, the initial neurological deficit was that of peripheral facial nerve palsy, even though a careful neurological examination diagnosed the patient as having a Bell's palsy. Facial palsy progressing slowly over a period of weeks or months, unremitting in its course and not attributable to another etiology, suggests involvement of a neoplasm [4–6].

Brain stem glioma usually presents with one or more cranial nerve palsies, cerebellar and pyramidal signs, and without any evidence of raised intracranial pressure [7–10]. The most common initial cranial nerve involvement is the abducens nerve rather than the facial nerve [7,10,11]. With tumor growth, however, the facial nerve may become involved because the abducens nucleus has a close anatomical relationship with the facial nerve. Therefore, brain stem tumors, in addition to facial nerve findings, are accompanied usually by more extensive neurological symptomatology. Isolated peripheral facial nerve palsy as an initial symptom is extremely rare. It is conceivable that, in our case, the tumor grew in the rostrocaudal direction along the nerve fibers [12] and did not involve other nuclei and long tracts.

Facial hyperkinesis, particularly in association with progressive palsy, also suggests that the problem is not idiopathic in origin [2]. Arseni and Petrovici [13] reported two patients with brain stem tumor who exhibited persistent tonic facial spasm with signs of gradual paresis. In our case, hyperlacrimation might be a sign of nervus intermedius irritation. It must be noted, however, that the typical clinical course of facial palsy due to neoplastic lesions is progressive.

Neuroradiological examination such as the MRI provides significant advantage in the diagnosis of CNS lesions. As noted in our case, MRI clearly demonstrated an intrinsic brain stem lesion. Until recently, the radiological diagnosis of Bell's palsy has been one of exclusion, arrived at only when imaging studies are normal. Recent neuroradiological reports [14,15] demonstrating enhancement of the intratemporal portion of the facial nerve on gadopentetate-dimeglumine-enahnced MRI in patients with Bell's palsy suggest that radiographic diagnosis of this entity may be possible rather than being inferred by an absence of abnormal findings. This anatomical pattern of facial nerve enhancement is consistent with the hypothesis that Bell's palsy results from viral geniculate ganglionitis with neural entrapment due to swelling within the tight confines of the fallopian canal. Contrast-enhanced MRI is recommended even in the evaluation of patients with isolated peripheral facial nerve palsy not only for exclusion of a CNS lesion, but also for positive diagnosis of Bell's palsy.

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