NOTEWORTHY CASES

ISOLATED FACIAL PALSY FROM PERINEURAL SPREAD OF CUTANEOUS SQUAMOUS CELL CARCINOMA

We evaluated an 82-year-old man with a 3-week history of acute onset of excessive tearing and mild difficulty closing the right eye. Past medical history included a squamous cell carcinoma of the right pre-auricular region, which had been excised 2 years earlier (1.8 cm in diameter, tumor-free margins). He subsequently underwent regular follow-up visits without evidence of recurrence.

Neurologic examination revealed mild weakness of the right orbicularis oculi and frontalis muscles. General examination was unremarkable, including skin inspection and palpation of cervical and submandibular lymph nodes and of the right parotid gland. Needle electromyography (EMG) demonstrated prominent fibrillation potentials and positive sharp waves in the territory of the temporal branch of the right facial nerve (orbicularis oculi and frontalis muscles), with markedly decreased recruitment in those muscles. EMG of the right orbicularis oris and mentalis muscles was normal. A 3-Tesla contrast-enhanced MRI scan of brain, temporal bones, face, and neck was normal.

Over the next 8 weeks, the patient's right facial palsy (FP) became complete (grade VI on the House-Brackmann scale). He also reported sensory impairment in the territory of the right great auricular nerve.

Needle EMG demonstrated worsening, with prominent fibrillation potentials, positive sharp waves, and no recordable voluntary motor unit potentials in the right orbicularis oculi, frontalis, orbicularis oris, and mentalis muscles, but with sparing of the posterior auricular muscle, suggesting a possible extracranial lesion. A repeat contrast-enhanced MRI scan and a subsequent positron emission tomography (PET) scan were normal. The patient then underwent a transcervical transparotid incisional biopsy of the main trunk of the right facial nerve, of its temporal and zygomatic branches, and of the great auricular nerve. Histopathologic examination revealed extensive perineural invasion of the facial nerve and the great auricular nerve by a poorly differentiated squamous cell carcinoma (Fig. 1A and B).

The patient was offered a radical surgical treatment, but he declined. He was then treated by external beam radiotherapy. The FP was still complete 2 years after onset.

Perineural spread (PS) of head and neck skin cancer is well recognized even if rare (5% of cases), with squamous cell carcinoma being the most common type.[1] Cancer invades nerve sheaths and spreads in a contiguous fashion within the perineurium, without lymphatic or hematogenous spread. Spread of the tumor can be either centrifugal or centripetal, sometimes with apparent skip areas.[2,3] Once the tumor cells reach a ganglion, a branch point, or are close to a contiguous nerve, they can spread proximally, distally, or along the contiguous nerve.[2,3] Clinical evidence of such spread manifests as cranial neuropathy in 30%–40% of patients with pathologic evidence of PS[4]; the fifth and seventh cranial nerve are most commonly involved because of their extensive distribution, but virtually all cranial nerves can be invaded, including the oculomotor nerves and the lower cranial nerves.[4]

There are rare case reports in the literature[5,6] of an isolated FP as the sole presentation of PS from a previously excised head and neck cutaneous malignancy. In the absence of cutaneous signs of disease recurrence, these patients can be misdiagnosed as having Bell's palsy. Our case highlights some clinical clues of a neoplasm as the cause of an FP in a patient with a history of a previously excised head and neck skin lesion. The selective and partial involvement of individual branches of the facial nerve is suggestive of a neoplastic etiology, in contrast to the total facial nerve involvement typical of Bell palsy. A slowly progressive course or the lack of recovery[5,7,8] should also raise suspicion for a neoplasm. The progressive involvement of multiple cranial nerves mandates extensive investigations to rule out an underlying invasive disease. It has been suggested[9] that communicating branches between the facial nerve and the trigeminal divisions or the cervical plexus may allow the spread of carcinoma of the skin between the motor and sensory systems of the head and neck. Our case highlights the presence of well-described[9] anastomotic branches between the facial and the great auricular nerve.

Other factors may delay the clinical suspicion of PS as the cause of FP. The primary tumor may have been

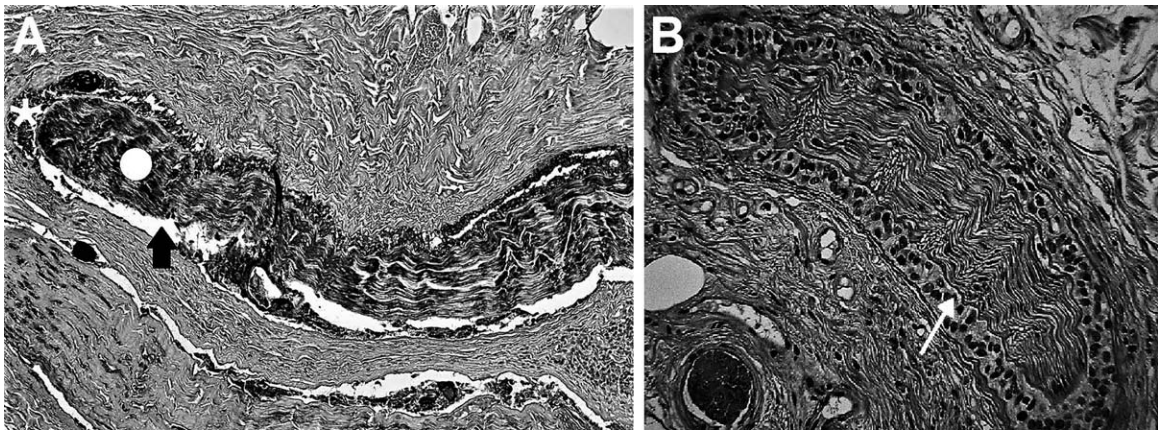

FIGURE 1. (A) Histologic specimen (light microscopy, 100 × magnification). Staining included hematoxylin–eosin and cytokeratin 19 (epithelial marker). The facial nerve (white dot) is cut longitudinally; perineural space (arrow) is the virtual empty space surrounding nerve fibers; tumor cells (white asterisk) are located within the perineural space. **(B)** High-power photomicrography (light microscopy, 200 × magnification, hematoxylin–eosin stain) shows cancer cells (arrow); the facial nerve is cut longitudinally.

excised long before (2 years in our case); in some cases[6] the patient may report an erroneous diagnosis of a benign skin lesion. Furthermore, there are often no skin lesions or reactive lymph node to detect on clinical examination, leading to the failure of clinicians to elicit a history of cutaneous tumor.

Some investigators[5] recommend imaging studies at 4 months from presentation with an FP if the clinical course is atypical for Bell palsy. The most sensitive radiologic method for the detection of PS from head and neck tumors is gadolinium-enhanced MRI, which has a sensitivity of 95%,[6,10] and should include brain, temporal bone, and parotid gland.[5] High-resolution CT scanning is a complementary technique for bone details. The role of PET scan in the diagnosis of perineural spread is unclear and its use in cutaneous malignancies has not been investigated thoroughly. Follow-up scans are necessary in patients whose initial investigations were negative but who have worsened[5]; if repeated imaging is negative and there is a high clinical suspicion of PS, facial nerve exploration and nerve biopsy of affected branches is suggested.[5] Biopsy must be extensive due to the possibility of skip areas; a repeated biopsy may be necessary if histology is negative and there is a high level of suspicion for malignancy.

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