Peripheral Facial Palsy Caused By Neoplastic Meningitis

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OBJECTIVES/HYPOTHESIS: To describe the clinical presentation of peripheral facial palsy caused by neoplastic meningitis.

STUDY DESIGN: Retrospective case series.

METHODS: Retrospective review, including accompanying symptoms and magnetic resonance imaging (MRI) findings.

RESULTS: Between January and December 2011, six patients were diagnosed with peripheral facial palsy caused by neoplastic meningitis. The patient age at presentation ranged from 56 to 77 years. The tumor origins were as follows: malignant lymphoma (n = 3), lung cancer (n = 2), and kidney cancer (n = 1). In three patients, sudden sensorineural hearing loss accompanied the facial palsy. Three patients were judged to be tumor-free at the onset of the facial palsy. In two patients, the malignancy was not diagnosed at the onset of facial palsy and hearing loss. Only one patient was diagnosed as having a tumor at the onset of the facial palsy. On the MRI, a mass lesion was detected in the internal auditory canals (IAC) of four patients; one had a solitary IAC tumor. In this patient, the diagnosis required a histopathological study using a translabyrinthine approach. In the other two patients, the facial nerve was enhanced with gadolinium, but it was difficult to differentiate the observed condition from Bell’s palsy. In these patients, repeated cytological studies led to the correct diagnoses.

CONCLUSIONS: The clinical symptoms and MRI findings of peripheral facial palsy caused by neoplastic meningitis sometimes mimic those of benign facial palsy.

KEY WORDS: Leptomeningeal carcinomatosis, carcinomatous meningitis, internal auditory canal metastasis, Bell’s palsy.

Level of Evidence: 4.

Laryngoscope, 124:2139–2143, 2014

INTRODUCTION

Peripheral facial palsy is a relatively common disease with diverse origins. The most frequent causes, Bell’s palsy and infectious disease, account for 65% to 90% of cases. Therefore, peripheral facial palsy is often diagnosed as Bell’s palsy or Ramsay Hunt syndrome when the ear drum and the external auditory canal are normal, the parotid gland tumor is not palpated, and cranial nerve functions are normal except for the facial and auditory nerve. However, we always have to keep it in mind that “All that palsies is not Bell’s.”

Peripheral facial palsies caused by tumors account for only 5% of all facial palsy cases, and most of these tumors are benign—including vestibular schwannomas, parotid tumors, cholesteatomas, and glomus jugulare tumors. Although facial palsies caused by malignant tumors are rare, they are clinically important because the prognosis differs greatly from that of the other causative conditions. Malignant tumors may violate the facial nerve anywhere from the parotid gland to the facial nucleus. The most common malignant tumor causing facial palsy is parotid cancer, and the importance of carefully reviewing the parotid gland is well recognized in the assessment of facial palsy. However, other malignant tumors are rarely recognized as a cause of facial palsy.

Neoplastic meningitis is a condition in which malignant tumor cells infiltrate into the meninges. It is also called as leptomeningeal carcinomatosis or carcinomatous meningitis. Neoplastic meningitis commonly shows tumor growth within the internal auditory canal (IAC) and can cause peripheral facial palsy (internal auditory canal metastasis). Although the incidence of neoplastic meningitis has been steadily rising, neoplastic meningitis has been underestimated as a cause of facial palsy, and few studies have reported about the clinical presentation of this condition.

In this article, we describe the clinical presentation of peripheral facial palsy caused by neoplastic meningitis.

MATERIALS AND METHODS

Between January 2011 and December 2011, 64 patients with peripheral facial palsy were studied. This study was supported by a Japan Society for the Promotion of Science (JSPS) Grant in Aid for Scientific Research (KAKENHI) Number 25462636 and the Japan Health Foundation. The authors have no other funding, financial relationships, or conflicts of interest to disclose.


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DOI: 10.1002/lary.24687
was conducted for hearing assessment. All patients received temporal bone computed tomography (CT) for the examination of the temporal bone lesion. Further exploration was conducted in patients with bilateral simultaneous palsy, involvement of multiple cranial nerves, signs of central nervous system lesion, concomitant malignancy, or no sign of recovery 3 months after the onset. Magnetic resonance imaging (MRI) with gadolinium enhancement was indicated for these patients. Patients were followed up until they had total recovery or were stable for more than 6 months. Forty-one cases were caused by Bell’s palsy, 11 cases were caused by Ramsay Hunt syndrome, three cases were caused by surgery, two cases were caused by parotid tumors, and one case was caused by otitis media. The remaining six patients were diagnosed with neoplastic meningitis. The medical records of these patients were retrospectively reviewed to collect the clinical information and the MRI findings.

In addition, we reviewed previous studies reported between 2001 and 2011 to assess the clinical and radiological findings of facial palsy caused by neoplastic meningitis.

RESULTS

Case 1
A 71-year-old male noticed dizziness 3 months before presenting at our institution. One month before presenting, he experienced sudden right ear sensorineural hearing loss and consulted another hospital. An MRI showed a solitary tumor in the right IAC (Fig. 1), and the patient was referred to our hospital with a diagnosis of vestibular schwannoma. In our department, he presented with mild right-side facial palsy (House-Brackmann grade 2). His hearing threshold was 65.0 dBHL in the right ear and 16.7 dBHL in the left ear. The patient did not demonstrate any other neurological abnormal findings. Six months prior to presenting, he underwent total resection of renal cell carcinoma (T3N0M0), following which there was no evidence of disease. His facial palsy deteriorated to grade 4 within 2 weeks. Surgical intervention was conducted via the translabyrinthine approach. The tumor was hemorrhagic and had invaded the facial nerve. The frozen section histology showed that the tumor was metastatic. Bony decompression with partial tumor resection was conducted. After surgery, the facial palsy improved to grade 2. Despite radiation and chemotherapy, the patient died of disease 4 months postsurgery.

Case 2
A 76-year-old male developed left-side facial palsy. Three days prior to the palsy, he noticed hearing loss in the left ear. He was diagnosed with small cell lung cancer, and brain metastasis was observed. The facial palsy was grade 3. His hearing threshold was 55.0 dBHL in the right ear and 77.5 dBHL in the left ear. The brain MRI revealed a tumor in the left IAC—in addition to disseminated tumors. He was administered steroids and underwent chemotherapy. His facial palsy deteriorated to grade 6, and hearing was eventually lost in both ears. The patient died of disease 7 months after the onset of the facial palsy.

Case 3
A 76-year-old male developed left-side facial palsy. He had been treated for malignant lymphoma of the testis 1 year prior and was deemed to be in complete remission. He was diagnosed with Bell’s palsy and was administered systemic steroids. Three months later, he developed grade 3 right-side facial palsy that was resistant to steroid treatment. He did not complain of hearing loss. The MRI showed enhancement along the facial nerve. Cytological study of cerebrospinal fluid revealed recurrence of the lymphoma. The right-side facial palsy recovered after chemotherapy. The patient died of disease 14 months after the onset of facial palsy.

Case 4
A 60-year-old female developed left-side facial palsy with ear pain. She had been treated for primary central nervous system lymphoma 8 months prior and was deemed to be in complete remission. She was diagnosed with Ramsay Hunt syndrome and was treated with steroids and antiviral drugs. Fourteen days later, she developed right-side facial palsy and consulted our hospital. The palsy was grade 6 on both sides. Pure tone audiometry did not show hearing loss. The patient complained of bilateral severe ear pain but did not show any other abnormal neurological findings. The MRI showed
enhancement along the facial nerve (Fig. 2). Repeated cytological study of the cerebrospinal fluid revealed recurrence of the lymphoma. The patient underwent chemo-radiotherapy. The facial palsy recovered to grade 1 on both sides after the treatment. The patient died of disease 30 months after the palsy.

Case 5
A 60-year-old female developed right-side facial palsy. She had no history of malignant tumors. The palsy was grade 6. Pure tone audiometry was normal. The patient did not show any other abnormal neurological findings and was diagnosed with Bell’s palsy. Two months later, she developed hypoesthesia of the left fingers and mild dementia. The MRI showed multiple tumors within the cranium, including the right IAC (Fig. 3). Systemic exploration was conducted, and the patient was diagnosed with lung cancer. The patient underwent chemoradiotherapy. She also underwent a static plastic surgery. The patient is alive with disease 27 months after the palsy.

Case 6
A 56-year-old male noticed hearing loss in the left ear and dizziness 2 weeks prior to presenting. He was diagnosed with idiopathic sudden sensorineural hearing loss and underwent steroid therapy. Subsequent MRI showed a brain tumor, and the patient was referred to our hospital. He was drowsy when he arrived at our department. The patient also had left hemiparesis and grade 2 right-side facial palsy. His hearing threshold was 1.7 dBHL in the right ear and 68.3 dBHL in the left ear. The following day, the patient’s facial palsy deteriorated to grade 3. After a biopsy, the patient was diagnosed with primary central nervous system lymphoma. He underwent chemotherapy. The facial palsy improved temporarily. The patient died of disease 7 months after the palsy.

Between 2001 and 2011, we found 16 published case reports of neoplastic meningitis causing facial palsy. A total of 22 cases, including the six cases from our series, were reviewed. The primary tumor site, initial symptoms, and brain MRI findings were summarized in Table 1. The most frequent accompanying symptom was hearing loss, followed by pain in the head and neck region. Fourteen patients had hearing loss; and five patients complained of ear or head pain around the time of onset of facial palsy. Brain MRI was performed in 21 cases. Five patients showed neuritis-like or normal brains. In eight cases, solitary tumors were found in the IAC and the cerebellopontine angle. Only eight cases showed multiple brain tumors or leptomeningeal enhancement. At the onset of peripheral facial palsy, a malignant tumor was suspected in only six cases. Five patients were initially diagnosed with Bell’s palsy or neuritis. In eight cases, benign IAC tumor was suspected.

**DISCUSSION**
In the diagnosis of acute facial palsy, a thorough neurological examination, ear examination, parotid gland palpation, and pure tone audiometry are routinely conducted. Abnormal findings other than facial palsy are an indication for imaging. Even with these workups, the underlying pathology can be overlooked. For example, both CT and MRI may not identify a small parotid gland tumor, especially if it is located in the deep lobe or close to the stylomastoid foramen. In our case series, three of six patients showed sudden sensorineural hearing loss occurring around the onset of the facial palsy, and two patients had concurrent malaise. The patients underwent MRI, and IAC tumors with/without intracranial dissemination were found. The other three patients had no abnormal findings other than peripheral facial palsy; they were initially diagnosed with Bell’s palsy or Ramsay Hunt syndrome. MRI was performed only after the emergence of new neurological symptoms: bilateral facial palsy, hypoesthesia of the left fingers, and mild dementia. In 1997, Imamura et al. reviewed the literature and summarized nine cases of neoplastic meningitis invading the temporal bone. The authors found that eight patients showed facial palsy. All nine patients had hearing loss, which was bilateral in seven cases. In most cases, the initial symptom was hearing loss; facial palsy developed later. Thus, the authors observed that the characteristic symptom of neoplastic meningitis is bilateral hearing loss followed by facial palsy. In recently published case reports, however, the clinical presentations seem different from those reviewed by Imamura et al. Among the 16 published cases reported between 2001 and 2011, five patients did not have hearing loss. Bilateral hearing loss was found in only two cases. In our series, hearing loss was found in three of six cases, and only one patient developed bilateral hearing loss—indicating that the clinical presentation of neoplastic meningitis has changed. This change may be partially attributed to early diagnosis of this condition. Goyault et al. reported the MRI findings of 14 patients with neoplastic meningitis involving the IAC. Among the 14 patients, only five showed sensorineural hearing loss. In three patients with hearing loss, the inner ear was enhanced with gadolinium. The authors speculated that...
the hearing loss did not occur before the tumor infiltrated the inner ear. Recent improvements in diagnostic tools may have allowed neoplastic meningitis involving the IAC to be diagnosed before it causes hearing loss.

MRI with gadolinium enhancement is the most useful imaging tool in patients with facial palsy suspected of having neoplastic meningitis. The most frequent MRI findings are subarachnoid nodules and leptomeningeal enhancement. Brain parenchymal metastases also indicate the neoplastic meningitis. However, MRI sensitivity is limited, particularly in patients with neoplastic meningitis caused by lymphoma and leukemia. The MRI sensitivity is reported to be as low as 29% to 46% in patients with hematological malignancies. In addition, the positive findings in MRI do not always lead to the correct diagnosis of neoplastic meningitis when the enhancement of the subarachnoid nodule is limited in the IAC. For example, in cases with a solitary IAC lesion, the lesion may be diagnosed as a vestibular schwannoma. In some cases—as in our case numbers 3 and 4—facial nerve enhancement mimicking the findings of Bell’s palsy or Ramsay Hunt syndrome is observed. In fact, only eight of the 22 reviewed cases were diagnosed as neoplastic meningitis at the time of the first MRI. In such cases, cytological or histological examination should be considered when the clinical findings suggest neoplastic meningitis.

Neoplastic meningitis has received little attention as a potential cause of peripheral facial palsy. However, this oversight appears to be changing. Neoplastic meningitis occurs in approximately 5% to 10% of all patients with malignant tumors, and the incidence has been steadily rising. Despite the high incidence, the clinical importance of neoplastic meningitis has been ignored because it usually occurs in the terminal stage of the malignant tumor. Recent improvements in chemotherapy have enabled the long-term survival of patients with systemic metastasis. Because chemotherapeutic agents have poor meningeal penetration, we are more likely to encounter neoplastic meningitis patients with good control of tumor at other sites. To diagnose neoplastic meningitis-derived facial palsy, regular checkups for neurological symptoms are needed during follow-up.

### Table 1: Literature for Peripheral Facial Palsy Caused by Neoplastic Meningitis

<table>
<thead>
<tr>
<th>Author</th>
<th>Primary Site</th>
<th>Palsy</th>
<th>Hearing loss</th>
<th>Initial Symptoms and Findings</th>
<th>Initial Diagnosis</th>
<th>Brain MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marques et al.</td>
<td>breast</td>
<td>unilateral</td>
<td>unilateral</td>
<td>neuritis</td>
<td>neuritis-like</td>
<td></td>
</tr>
<tr>
<td>Martin-Duverneuil et al.</td>
<td>lung</td>
<td>unilateral</td>
<td>unilateral</td>
<td>–</td>
<td>IAC tumor</td>
<td></td>
</tr>
<tr>
<td>Guilemany et al.</td>
<td>breast</td>
<td>unilateral</td>
<td>unilateral</td>
<td>VS</td>
<td>CPA/IAC tumor</td>
<td></td>
</tr>
<tr>
<td>Lee et al.</td>
<td>nasopharynx</td>
<td>unilateral</td>
<td>–</td>
<td>Anemia, facial numbness, facial pain</td>
<td>–</td>
<td>leptomeningitis</td>
</tr>
<tr>
<td>Baba et al.</td>
<td>stomach</td>
<td>unilateral</td>
<td>bilateral</td>
<td>headache</td>
<td>Ramsay Hunt</td>
<td>normal brain</td>
</tr>
<tr>
<td>Schrock et al.</td>
<td>lung</td>
<td>unilateral</td>
<td>unilateral</td>
<td>VS</td>
<td>IAC tumor</td>
<td></td>
</tr>
<tr>
<td>Brackmann and Doherty</td>
<td>MM</td>
<td>unilateral</td>
<td>unilateral</td>
<td>Facial hypoesthesia, headache, middle ear mass</td>
<td>facial schwannoma</td>
<td>CPA/IAC tumor</td>
</tr>
<tr>
<td>Chang et al.</td>
<td>lung</td>
<td>unilateral</td>
<td>unilateral</td>
<td>malignancy</td>
<td>CPA/IAC tumor</td>
<td></td>
</tr>
<tr>
<td>Favier et al.</td>
<td>unknown</td>
<td>bilateral</td>
<td>unilateral</td>
<td>VS</td>
<td>IAC tumor</td>
<td></td>
</tr>
<tr>
<td>Kariya et al.</td>
<td>RMS</td>
<td>unilateral</td>
<td>bilateral</td>
<td>Hoarseness, dysphagia, stiff neck</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>Kubo et al.</td>
<td>lung</td>
<td>unilateral</td>
<td>–</td>
<td>metastasis</td>
<td>multiple tumor*</td>
<td></td>
</tr>
<tr>
<td>Chotani et al.</td>
<td>PNET</td>
<td>unilateral</td>
<td>–</td>
<td>facial hotness, ear pain</td>
<td>neurofibromatosis</td>
<td>multiple tumor</td>
</tr>
<tr>
<td>Della Puppa et al.</td>
<td>breast</td>
<td>unilateral</td>
<td>unilateral</td>
<td>metastasis</td>
<td>multiple tumor</td>
<td></td>
</tr>
<tr>
<td>Quadri et al.</td>
<td>ML</td>
<td>unilateral</td>
<td>–</td>
<td>abducent paresis, neck pain, limb palsy</td>
<td>neurofibromatosis</td>
<td>normal brain</td>
</tr>
<tr>
<td>Yildiz et al.</td>
<td>lung</td>
<td>unilateral</td>
<td>–</td>
<td>facial numbness, dysphonia, weight loss, lymphadenopathy</td>
<td>metastasis</td>
<td>multiple tumor</td>
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<tr>
<td>Present study</td>
<td>kidney</td>
<td>unilateral</td>
<td>unilateral</td>
<td>VS</td>
<td>CPA/IAC tumor</td>
<td></td>
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<tr>
<td></td>
<td>lung</td>
<td>unilateral</td>
<td>bilateral</td>
<td>malaise</td>
<td>metastasis</td>
<td>multiple tumor</td>
</tr>
<tr>
<td></td>
<td>ML</td>
<td>bilateral</td>
<td>–</td>
<td>Bell’s palsy</td>
<td>Bell’s palsy</td>
<td>neuritis-like</td>
</tr>
<tr>
<td></td>
<td>ML</td>
<td>bilateral</td>
<td>–</td>
<td>Ramsay Hunt</td>
<td>multiple tumor</td>
<td></td>
</tr>
<tr>
<td></td>
<td>lung</td>
<td>unilateral</td>
<td>–</td>
<td>Bell’s palsy</td>
<td>malignancy</td>
<td>multiple tumor</td>
</tr>
<tr>
<td></td>
<td>ML</td>
<td>unilateral</td>
<td>malaise</td>
<td>metastasis</td>
<td>CPA/IAC tumor</td>
<td></td>
</tr>
</tbody>
</table>

*Bilateral hearing loss occurred at the terminal stage.*

CPA = cerebellopontine angle; IAC = internal auditory canal; ML = malignant lymphoma; MM = malignant melanoma; PNET = primitive neuroectodermal tumor; RMS = rhabdomyosarcoma; VS = vestibular schwannoma.
CONCLUSION
Neoplastic meningitis can be a cause of peripheral facial paralysis in patients with apparently good general health. The clinical symptoms are sometimes nonspecific. The most frequent accompanying symptom is hearing loss, but this symptom is not always present. Careful follow-up is necessary to detect delayed abnormal neurological symptoms. Patients with suspected neoplastic meningitis should be subjected to imaging. MRI with gadolinium enhancement is the most useful imaging tool. Nevertheless, the MRI findings may mimic those of Bell’s palsy and vestibular schwannoma. In such cases, cytological or histological examination is indicated.

BIBLIOGRAPHY