Case report

Bilateral peripheral facial palsy and mastoid infiltration as symptoms of relapsed acute myeloid leukemia

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ABSTRACT

Background: Although Bell’s palsy (BP) is the most common cause of peripheral facial palsy (PFP), other etiologies merit investigation.

Case report: A 60-year-old female patient presented with recurrent bilateral PFP. Although the patient had a history of acute myeloid leukemia (AML), she had initially been diagnosed with BP-related PFP and had been treated accordingly. When the PFP recurred, additional diagnostic tests were performed. The resulting immunohistochemical profile included CD3 positivity in a few reactive T lymphocytes; positivity for myeloperoxidase in atypical cells; and focal positivity for CD34 and proto-oncogene c-kit proteins in neoplastic cells, thus confirming the suspicion of mastoid infiltration caused by relapsed AML.

Conclusion: In patients with neoplastic disease, a finding of PFP calls for extensive investigation in order to rule out the involvement of the temporal bone.

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1. Introduction

Bell’s palsy (BP) is the most common cause of peripheral facial palsy (PFP) and has the lowest rate of sequelae; the incidence of BP is low (20–30/100,000 population) and complete remission of symptoms occurs in 83% of cases [1]. Because other etiologies of PFP are rarely suspected or investigated, it is possible that the role of BP as a cause of PFP is overestimated. Among patients with PFP, the unilateral form is more common than is the bilateral form, which is seen in only 0.3–2.0% of such patients [2]. Bilateral PFP is more frequently associated with systemic causes, and ancillary tests should therefore be performed before a diagnosis of bilateral PFP is made [2].

We report the case of an adult patient in whom the etiology of unilateral PFP was incorrectly identified as BP. That assumption was proved incorrect after the patient presented with recurrence of the PFP (bilateral in this second instance) and mastoid infiltration (a rare finding), both of which were determined to be symptoms of relapsed acute myeloid leukemia (AML). We emphasize the need for a detailed differential diagnosis, in order to rule out alternative etiologies, in similar cases.

2. Case report

A 60-year-old female patient sought medical attention in our department of otolaryngology and head and neck surgery, presenting with a seven-day history of right-sided PFP. The patient complained of non-specific bilateral otalgia. She reported no dizziness, tinnitus, otorrhea, or fever. The patient had a history of AML, which had been treated with induction chemotherapy with cytarabine and daunorubicin, followed by allogeneic hematopoietic stem cell transplantation and radiation therapy in the paravertebral region 70 days prior. Physical examination revealed House-Brackmann grade III PFP and the absence of Bell’s phenomenon. Otoscopic examination findings were normal. Audiometry revealed bilateral sensorineural hearing loss of up to 40 dB at 4 kHz, 6 kHz, and 8 kHz, as well as showing a type A tympanogram and normal stapedial reflexes in both ears. On the basis of a presumptive diagnosis of BP, the patient was treated with oral corticosteroids and acyclovir. Subsequently, there was complete resolution of the symptoms. Forty-five days later, the patient again presented with PFP but had no other complaints. Physical examination revealed House-Brackmann grade IV right-sided PFP and bulging of the posterior wall of the external auditory canal, as well as ipsilateral hyperemia and purulent discharge preventing visualization of the tympanic membrane. There were signs of otitis media with effusion in the left ear, and bilateral mixed hearing loss was confirmed by audiometry. One day later, the patient developed House-Brackmann grade III left-sided PFP.

A computed tomography scan of the mastoids was inconclusive, showing bilateral opacification with increased soft tissue density...
Fig. 1. In A, coronal computed tomography (CT) scan of the middle ear and right mastoid. In B, CT scan showing dehiscence of the right facial canal. In C, sagittal T1-weighted magnetic resonance image showing lateral sinus thrombosis.

Fig. 2. In A, mastoid bone fragments interspersed with immature myeloid cells (hematoxylin and eosin [H&E] staining; magnification, ×40). In B, mastoid mucosa fragments interspersed with immature myeloid cells (H&E staining; magnification, ×10). In C, focal positivity for proto-oncogene proteins c-kit in neoplastic cells (immunohistochemistry; magnification, ×40). In D, positivity for myeloperoxidase in atypical cells (immunohistochemistry; magnification, ×40).
Although there were no areas of bone destruction, there was bilateral dehiscence of the facial canal in the tympanic portion of the facial nerve. Cranial magnetic resonance imaging showed signs of sigmoid sinus thrombosis on both sides.

The fluid obtained through lumbar and right tympanic membrane punctures showed no neoplastic cells or microorganisms. Analysis of the peripheral blood revealed pancytopenia and no blast cells.

To collect material for analysis, we performed right antrostomy (of the mastoid antrum), with the patient under general anesthesia. Throughout the procedure, continuous and profuse bleeding occurred when the cortical bone was drilled. The mastoid antrum contained no soft tissue. Examination of the material collected revealed blasts amid the mastoid bone fragments, a finding that was at odds with the absence of immature cells in the peripheral blood (Fig. 2A and B). The immunohistochimical profile was as follows: negativity for CD20; positivity for CD3 in a few reactive T lymphocytes; positivity for myeloperoxidase in atypical cells (Fig. 2D); and focal positivity for CD34 and proto-oncogene proteins c-kit in neoplastic cells (Fig. 2C). Those findings supported the diagnosis of mastoid infiltration due to AML. After the surgical procedure, there was a progressive improvement in the PFP. After the patient had been diagnosed with relapsed AML, chemotherapy was resumed. However, the patient developed febrile neutropenia and sepsis, resulting in her death at 13 days after the initiation of treatment.

3. Discussion

Because BP is the most common cause of PFP and because the rates of remission of BP are high, many PFP cases in which there is complete resolution of symptoms are misdiagnosed as cases of BP, despite the fact that all other possible causes of PFP should be ruled out before a diagnosis of BP can be made [3]. Although there is an extensive list of differential diagnoses for recurrent or bilateral PFP, the etiology remains unknown in most patients [4]. The major systemic causes of PFP include Guillain–Barre syndrome, cranial nerve diseases, Melkersson–Rosenthal syndrome, brainstem encephalitis, benign intracranial hypertension, syphilis, leukemia, sarcoidosis, Lyme disease, meningitis, and acute HIV infection.

Leukemia is a malignant disease characterized by abnormal proliferation of white blood cells and their precursors. Symptomatic otologic involvement by leukocyte infiltration is unusual, most often occurring in patients previously diagnosed with leukemia [5]. In 2002, Rhee et al. suggested that recurrent PFP is associated with relapsed leukemia [6]. Likewise, Buyukavci et al. reported that bilateral PFP is a warning sign of leukocyte infiltration [7]. In the case reported here, the recurrence of PFP and the fact that the patient presented with bilateral PFP prompted a more detailed investigation.

A few cases of PFP due to AML have been reported in children [8,9]. However, this is the first histologically confirmed case to be reported in an adult in the last 20 years.

Although we found facial canal erosion in our patient, neoplastic cell invasion of the facial canal does not necessarily cause PFP, which is believed to occur only when such cells cross the epineural sheath. Even in such cases, complete remission of PFP and complete recovery of facial function can be achieved with treatment [10].

4. Conclusion

In conclusion, the occurrence of PFP in patients with neoplastic disease (hematological or otherwise) requires extensive investigation in order to rule out temporal bone involvement.

Disclosure of interest

The authors have not supplied their declaration of conflict of interest.

References