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Case Reports

Facial nerve paresis as the presenting symptom of leukemia *

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Abstract

Leukemic involvement of the temporal bone is not uncommon and may present in a variety of ways including auricular or external canal skin lesions, red or thick tympanic membrane, middle ear effusions, otitis media, hearing loss or mastoiditis. Symptomatic facial nerve involvement, on the other hand, is extremely unusual. We discuss a pediatric patient whose sudden onset facial nerve paresis was the presenting symptom that led to her diagnosis of leukemia. At the time of mastoidectomy, a granulocytic sarcoma or chloroma was noted to be overlying the VIIth nerve.

Introduction

Leukemia is a malignant disease of the blood-forming organs that is characterized by distorted proliferation and development of leukocytes and their precursors. It can affect virtually any part of the body either directly (i.e. leukemic infiltrates) or indirectly as a result of abnormal hematopoietic function (i.e. infection, hemorrhage). The head and neck are frequently involved in the leukemic process, the ear less so. Gotay [2] noted that leukemia of the temporal bone may present to the otolaryngologist as skin lesions of the auricle or external auditory canal, red or thick tympanic membrane, hemorrhages or exudates in the middle ear,

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acute otitis media, conductive or sensorineural hearing loss, acute mastoiditis or facial paralysis.

Zechner and Altmann [6], in their review, noted that 32/100 previously reported leukemic patients had otologic symptoms, with hearing loss being the most common complaint. In 6 of these patients, the otologic problem was their presenting complaint. Of the 14 new patients presented, 3 had symptomatic otologic problems (mixed hearing loss, bloody otorrhea, infiltration of the tympanic membrane). In the majority of cases, the perineural, and to a lesser extent, the endoneural sheaths of the VIIth and VIIIth nerves, were diffusely infiltrated. In 2 of these patients, heavy tumor-like infiltrations of the horizontal portion of the facial nerve were noted, although none had VIIth nerve symptoms.

To date, Paparella et al. [4] have reported the largest series of temporal bones from leukemic patients. In their evaluation of the temporal bones of 25 leukemic patients they found that 48% of patients demonstrated clinical otologic problems, although a leukemic etiology on histologic evaluation could only be established in 20%. In 11 cases leukemic infiltrates of the perineural and, to a lesser extent, the endoneural sheaths of the VIIth and VIIIth nerves in the internal auditory meatus were observed. In 2 patients, the Fallopian canal was infiltrated with leukemic cells in the horizontal and vertical portions. In none of these patients was there clinical evidence of VIIth nerve dysfunction.

History

S.K. was a previously healthy 6-year-old white female who presented to her pediatrician with a cough and a temperature of 102° F. She was placed on a course of amoxicillin with defervescence and symptomatic improvement. On day 9 after initial presentation, she developed left otalgia and the following morning her mother noted facial asymmetry upon awakening. She was seen again by her pediatrician who suspected altered facial nerve function secondary to otitis media and referred immediately to the Otolaryngology Department at the University of Michigan.

Her past medical history was remarkable for 2 URIs during the previous winter and 2 episodes of otitis media—most recently 2 years prior to admission. There was a question of recent weight loss but she had been without illnesses other than those already mentioned. No fatigue, night sweats or unusual bruising had been noted.

She was afebrile and in no acute distress on presentation. Her left tympanic membrane was opaque and bulging while the right tympanic membrane was clear with a serous middle ear effusion present behind it. No mastoid tenderness or swelling was appreciated. Diffuse bilateral shotty posterior cervical adenopathy was present. Cranial nerves were intact except for a left facial paresis—III/VI (I/VI, normal function; VI/VI, complete paralysis) forehead and eye, VI/VI nose and IV/VI mouth (Fig. 1).

A complete blood count obtained upon admission revealed Hgb, 9.7; Hct, 28.2; platelets, 115,000; segs, 9; bands, 7; lymphs, 61; metamyelocytes, 3; myelocytes, 1; and myeloblasts, 19. A wide myringotomy was performed urgently. The left middle

Fig. 1. This photograph of S.K. at the time of presentation shows the left facial weakness.

ear cavity was noted to be filled with mucopurulent fluid and the Gram stain showed moderate WBCs but no organisms.

The following day a bone marrow aspirate was obtained and was consistent with the diagnosis of acute non-lymphocytic leukemia. A CAT scan showed opacification of the left mastoid air cells and middle ear cleft with considerable swelling of the middle ear mucosa (Fig. 2). The findings on the right were similar but less severe. In addition, on the left side there was a collection of opacification without trabeculation (consistent with a leukemic focus or chloroma) that closely approached the superior aspect of the Fallopian canal.

On the third day, while the patient was under general anesthesia for placement of a chemotherapy port, a biopsy of the middle ear mucosa was performed. The histopathology revealed a leukemic infiltrate involving the submucosa.

The facial paresis remained unchanged and, on the seventh hospital day, a left tympanotomy with mastoidectomy was performed. The hypotympanum and epitympanum were filled with a greenish gelatinous material that was noted to overlie the second genu (horizontal portion) of the VIIth nerve. There was no evidence of bony dehiscence along the Fallopian canal. The histopathology sections were consistent with granulocytic sarcoma or chloroma (Fig. 3).

Chemotherapy was begun the following day. None of the cultures grew any organisms. Facial function slowly returned and by the third month after diagnosis

Fig. 2. A CAT scan of the temporal bones shows the soft tissue density in the mastoid and middle ear cavities bilaterally. An area with loss of trabeculations can be seen in the left mastoid cavity (arrow) adjacent to the facial nerve. This is consistent with a soft tissue mass.

facial nerve function was normal except for II/VI function in the left eye distribution.

Discussion

A granulocytic sarcoma, or chloroma, is a localized accumulation or 'tumor' composed of immature cells of the granulocytic series [3,5]. The name chloroma was given in 1853 by King because the tumors often display a greenish color that fades with exposure to air. The color is due to myeloperoxidase in the malignant granulocytes and their precursors. The most common sites of involvement are the periosteum, soft tissues, bone, lymph nodes and skin [3].

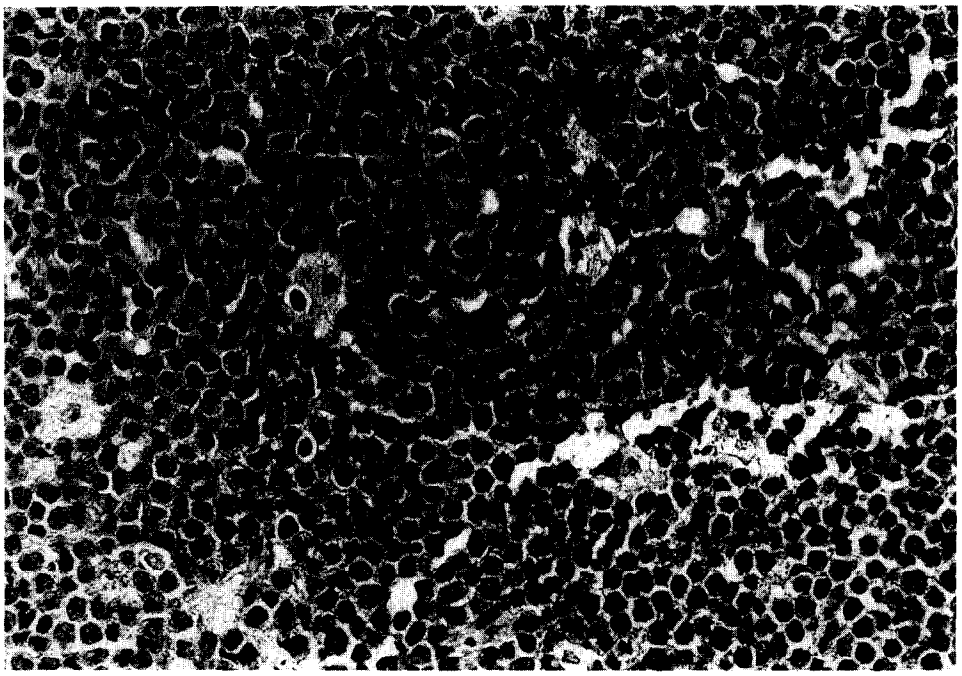

Fig. 3. A histologic section of the chloroma or granulocytic sarcoma demonstrating the sheets of immature granulocytes (hematoxylin-eosin, ×243).

Nieman et al. [3] evaluated the chloromas of 50 patients and noted that 24 of these patients had a known myeloproliferative disorder other than acute myeloid leukemia (most commonly chronic granulocytic leukemia). Eleven patients had chloroma either as the presenting feature that led immediately to the diagnosis of acute myeloid leukemia (AML) (6/11), or the chloroma was observed after the diagnosis of AML had been made (5/11). Fifteen patients had no known hematopoietic disease discovered at the time of presentation. Of these, 13/15 developed AML from 1–49 months afterwards. The remaining two patients died 3 and 10 months after discovery of the chloroma without evidence of hematologic abnormality. In our patient, the chloroma was present at the time of diagnosis of leukemia.

The purulent middle ear effusion in the patient presented here was unaccompanied by any other signs of otitis media. The tympanic membrane was bulging but erythema and tenderness with pneumotoscopy were absent. In 2 of 4 temporal bones of leukemic patients, Druss [1] noted extensive suppuration of the middle ear but neither patient had otologic symptoms. Paparella et al. [4] also noted leukemic infiltration of the middle ear. When the patients had complaints, the infiltrates were always noted to be moderate to severe, whereas when there were no symptoms, the infiltrate was mild to moderate.

In 1984, Todd and Bowman [5] reported a patient who presented with facial paralysis and mastoiditis. They performed a limited myringotomy without much

drainage of fluid or relief of symptoms. Four days later, a mastoidectomy revealed a tan rubbery lobulated mass filling the mastoid antrum and facial recess, encasing the ossicles and extending into the Eustachian tube. The diagnosis of acute myelogenous leukemia was made after a postoperative complete blood count (CBC) differential was performed and the final pathology report on the mass defined it as a chloroma.

Although unusual, the temporal bone manifestation may be the presenting symptom or sign of leukemia. The patient discussed in this paper presented with the extremely unusual symptom of facial nerve paresis and it was the CBC differential that led to the suspicion of leukemia. The chloroma overlying the VIIth nerve appeared to be the cause of the weakness.

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