RECURRENT UNILATERAL FACIAL NERVE PALSY AS AN ATYPICAL PRESENTATION OF ACUTE MYELOID LEUKEMIA: A DIAGNOSTIC CHALLENGE

PARALYSIE FACIALE UNILATÉRALE RÉCURRENTE COMME PRÉSENTATION ATYPIQUE DE LA LEUCÉMIE AIGUË MYÉLOÏDE: UN DÉFI DIAGNOSTIQUE

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ABSTRACT

Introduction: Acute promyelocytic leukemia (APL) relapse rarely presents with otologic manifestations. Facial nerve palsy associated with external auditory canal (EAC) lesions may mimic benign conditions such as cholesteatoma, leading to diagnostic delay.
Case report: We report the case of a 23-year-old male with a history of recurrent right peripheral facial nerve palsy and acute myeloid leukemia (AML M3) in remission for six years. He presented with a new episode of facial palsy associated with acute conductive hearing loss. Otoscopic examination revealed a bleeding, polypoid mass filling the EAC. Imaging suggested cholesteatoma with bone erosion and facial canal dehiscence. However, during hospitalization, systemic symptoms prompted hematological evaluation, revealing a relapse of APL with 70% promyelocytic blasts and PML-RARA positivity. Following induction chemotherapy, a marked regression of otologic lesions was observed.
Discussion: This case highlights an infrequent presenting symptom of AML relapse and a rare cause of recurrent facial nerve palsy. Leukemic infiltration may closely mimic cholesteatoma but requires no surgery: targeted induction therapy led to regression of the lesion and improvement of the facial palsy, while early control of the coagulopathy was potentially life-saving. Finally, recurrent facial palsy with atypical otologic findings warrants a hematological workup, particularly in patients with a history of leukemia.
Keywords: Acute myeloid leukemia; Recurrent facial palsy; Extramedullary relapse; Temporal bone involvement.

RÉSUMÉ

Introduction: La rechute d'une leucémie aiguë promyélocytaire (LAP) se manifeste rarement par des signes otologiques. Une paralysie faciale associée à des lésions du conduit auditif externe (CAE) peut simuler des affections bénignes telles qu'un cholestéatome, entraînant un retard diagnostique.
Cas clinique: Nous rapportons le cas d'un homme de 23 ans aux antécédents de paralysie faciale périphérique droite récidivante et de leucémie aiguë myéloïde (LAM M3) en rémission depuis six ans. Il s'est présenté pour un nouvel épisode de paralysie faciale associé à une surdité de transmission aiguë. L'examen otoscopique a révélé une masse polypoïde hémorragique comblant le conduit auditif externe. L'imagerie évoquait un cholestéatome avec érosion osseuse et déhiscence du canal facial. Cependant, au cours de l'hospitalisation, l'apparition de signes généraux a conduit à une évaluation hématologique, révélant une rechute de LAP avec 70 % de blastes promyélocytaires et une positivité PML-RARA. Après instauration d'une chimiothérapie d'induction, une régression marquée des lésions otologiques a été observée.
Discussion: Ce cas illustre un symptôme de rechute de LAM peu fréquent et une cause rare de paralysie faciale récidivante. L'infiltration leucémique peut fortement simuler un cholestéatome, mais ne nécessite pas d'intervention chirurgicale: un traitement d'induction ciblé a permis la régression de la lésion et l'amélioration de la paralysie faciale, tandis que la prise en charge précoce de la coagulopathie a potentiellement sauvé la vie du patient. Enfin, une paralysie faciale récidivante associée à des signes otologiques atypiques justifie un bilan hématologique, notamment chez les patients ayant des antécédents de leucémie.
Mots-clés: Leucémie aiguë myéloïde; Paralysie faciale récidivante; Rechute extramédullaire; Atteinte de l'os temporal.

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INTRODUCTION

Acute promyelocytic leukemia (APL), a distinct subtype of acute myeloid leukemia characterized by the t(15;17) translocation leading to the PML-RARA fusion gene, is currently associated with high cure rates exceeding 80% due to targeted therapies such as all-trans retinoic acid and arsenic trioxide. Despite these advances, relapse still occurs in a minority of patients and may present with atypical or extramedullary manifestations [1,2].

Extramedullary relapse of APL is uncommon but increasingly recognized, particularly in the central nervous system and, more rarely, in the head and neck region. Temporal bone involvement is exceptional and may present with nonspecific otologic symptoms, including hearing loss, otorrhea, or facial nerve palsy [3,4].

Peripheral facial nerve palsy is a rare manifestation of leukemia and may occur either at initial diagnosis or during relapse. Its pathophysiology involves leukemic infiltration of the facial nerve, meningeal involvement, or compression by granulocytic sarcoma [5,6]. Importantly, recurrent or atypical facial nerve palsy should raise suspicion for an underlying systemic disease, especially in patients with a history of hematologic malignancy [6].

Leukemic infiltration of the temporal bone may radiologically and clinically mimic benign otologic conditions such as cholesteatoma or chronic otitis media. Several reports have highlighted cases in which granulocytic sarcoma of the external auditory canal presented as a cholesteatoma-like lesion with bone erosion, leading to diagnostic confusion [7,8].

This overlap represents a significant diagnostic challenge for otolaryngologists and may result in delayed recognition of leukemia relapse. We present a rare case of recurrent facial nerve palsy revealing a late relapse of APL, initially misdiagnosed as external auditory canal cholesteatoma.

CASE PRESENTATION

A 23-year-old male presented with a one-month history of right-sided peripheral facial nerve palsy associated with acute-onset hypoacusis. His medical history was significant for recurrent right facial nerve palsy (four previous episodes) and acute myeloid leukemia (AML M3), treated in 2017 with induction chemotherapy followed by three consolidation courses (idarubicin plus all-trans retinoic acid [ATRA], mitoxantrone plus ATRA, and idarubicin plus ATRA), with completion of treatment after 2 years and complete remission achieved in 2019. The previous episodes of facial palsy each lasted approximately one week and were treated with corticosteroids, antiviral therapy and physiotherapy, with partial recovery (Freyss facial grading score of 17/30).

Clinical examination revealed residual facial nerve deficits. Otomicroscopy examination demonstrated a polypoid, friable, and bleeding lesion filling the external auditory canal, predominantly arising from the posterior wall.

Temporal bone computed tomography (CT) showed an epitympanic soft tissue lesion with convex margins extending into the posterior wall of the external auditory canal (EAC), associated with ossicular chain erosion, tegmen tympani destruction, and dehiscence of the facial nerve canal involving its second and third segments (Figure 1).

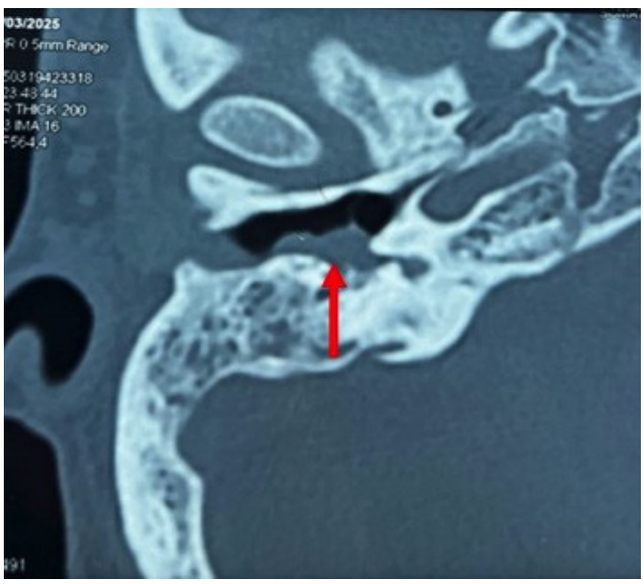


Figure 1: Petrous bone CT, axial section, showing polypoid thickening of the posterior wall of the EAC, without bone lysis or suspicious contrast enhancement.

Magnetic resonance imaging (MRI) suggested a diagnosis of right-sided cholesteatomatous otitis (Figure 2).



Figure 2: Contrast-enhanced T1-weighted MRI shows a poorly demarcated expansile process involving the right otomastoid region, occupying nearly the tympanic cavity and mastoid antrum, with extension into the ipsilateral EAC.

Pure-tone audiometry revealed a right-sided conductive hearing loss of 45 dB.

During hospitalization, the patient developed coxofemoral pain, diffuse ecchymotic lesions, and hematuria without fever. These findings prompted further hematological evaluation. Bone marrow examination revealed 70% blasts with promyelocytic morphology. Immunophenotyping and molecular analysis confirmed a relapse of APL with PML-RARA translocation, associated with hyperleukocytosis and disseminated intravascular coagulation.

The patient received induction chemotherapy in accordance with National Comprehensive Cancer Network (NCCN) guidelines for the management of APL, induction therapy was initiated based on a combination regimen of all-trans retinoic acid (ATRA), idarubicin (Ida), and arsenic trioxide (ATO) without the need for surgery. The outcome was marked by significant clinical improvement, including marked regression of the external auditory canal lesion and facial paralysis, as well as radiological improvement on follow-up MRI (Figure 3).

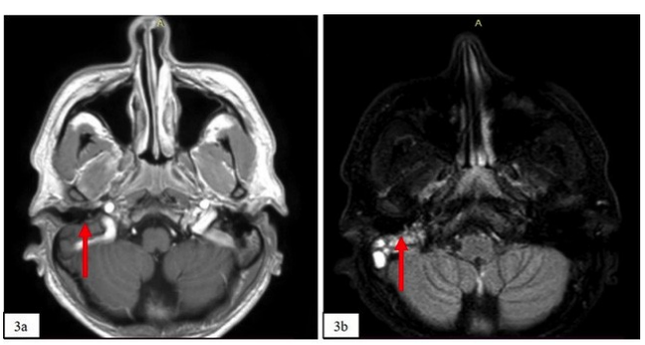


Figure 3 (a,b): Contrast-enhanced T1-weighted and T2-weighted MRI demonstrates regression of the soft-tissue filling of the ECA, with persistent marrow replacement involving the right mastoid and right occipital condyle.

DISCUSSION

This case illustrates a rare and diagnostically challenging presentation of APL relapse, emphasizing the importance of considering hematologic malignancy in atypical otologic presentations.

Facial nerve palsy in leukemia remains an uncommon but well-documented phenomenon. It may result from direct leukemic infiltration of the facial nerve, leukemic meningitis, or compression by extramedullary masses such as granulocytic sarcoma [5,6]. In our patient, the history of recurrent facial nerve palsy is particularly noteworthy, as repeated episodes should be considered a warning sign of an underlying systemic or neoplastic process rather than idiopathic Bell's palsy.

Several authors have reported that facial nerve palsy may represent an early or even the first manifestation of leukemic relapse, sometimes preceding hematological abnormalities [6,9]. This highlights the need for a high index of suspicion, particularly in patients with a prior history of leukemia.

Temporal bone involvement in leukemia is rare but increasingly recognized due to improved imaging techniques. The most common sites include the middle ear, mastoid, and less frequently the external auditory canal. Clinical manifestations are variable and may include conductive hearing loss, otalgia, otorrhea, and cranial nerve deficits [3,4,10].

Radiologically, leukemic infiltration may closely mimic cholesteatoma, as both entities can present with soft tissue masses and aggressive bone destruction, including ossicular erosion, tegmen tympani defects, and facial canal dehiscence. In our case, CT and MRI findings strongly suggested cholesteatoma, illustrating the limitations of imaging in differentiating these entities [7,8].

Granulocytic sarcoma, an extramedullary tumor composed of immature myeloid cells, represents a key differential diagnosis. It has been reported in the temporal bone and external auditory canal, where it may present as a polypoid, friable, and bleeding mass — features that were also observed in our patient [7]. Importantly, such lesions are frequently misdiagnosed as inflammatory or benign conditions, including cholesteatoma.

Histopathological examination is usually considered the gold standard for diagnosis. However, as demonstrated in this case, biopsy may be misleading, particularly when samples are superficial, necrotic, or contaminated by inflammatory tissue. This underlines the importance of clinicopathological correlation and the need for repeat or deeper biopsies when clinical suspicion persists.

A critical turning point in our case was the onset of systemic symptoms, including ecchymosis, hematuria, and bone pain, which prompted hematological reassessment. The diagnosis of APL relapse was confirmed by bone marrow examination and detection of the PML-RARA fusion gene. Notably, APL relapse is often associated with coagulopathy, including disseminated intravascular coagulation, which represents a life-threatening complication requiring urgent treatment [1,2].

The rapid regression of the otologic lesion following induction chemotherapy strongly supports its leukemic origin. This therapeutic response is a key diagnostic argument in favor of leukemic infiltration or granulocytic sarcoma rather than true cholesteatoma.

In line with our findings, previous reports have demonstrated that facial nerve palsy secondary to leukemic infiltration may significantly improve with chemotherapy alone, without the need for surgical intervention. In particular, Fana et al. described a case of acute myeloid leukemia presenting with facial nerve palsy associated with mastoid involvement, in which intrathecal chemotherapy (methotrexate and cytarabine) led to rapid clinical improvement of facial paralysis [11]. This observation supports the hypothesis that facial nerve dysfunction in leukemia is often related to inflammatory or infiltrative mechanisms that are highly

responsive to systemic or intrathecal chemotherapy rather than requiring surgical decompression.

Similar cases have been reported in the recent literature. Breccia et al. described seven patients with ear involvement at APL relapse, mainly presenting with hypoacusis and earache; two of them were in hematologic and molecular remission, suggesting that the ear may act as a "sanctuary" site [12]. Choi et al. reported a bleeding mass arising from the posterior wall of the external auditory canal in relapsed APL, which regressed with ATRA and arsenic trioxide without surgery, a presentation very close to that of our patient [8]. Leite da Silveira et al. described a patient with AML whose facial palsy was first treated as Bell's palsy and then recurred with mastoid infiltration by leukemic cells [13]. In a recent review of 14 cases of temporal bone myeloid sarcoma, facial asymmetry was among the most frequent symptoms, and most patients had a history of AML [14].

These observations emphasize the need for a comprehensive approach in patients with recurrent neurological symptoms. Bell's palsy remains a diagnosis of exclusion, and current guidelines recommend reassessment when the course is atypical [15]. A recurrent ipsilateral facial palsy should therefore prompt temporal bone imaging and a hematological workup, including a complete blood count with peripheral smear and coagulation tests, and, in patients with a history of leukemia, bone marrow examination and molecular monitoring (PML-RARA).

From a clinical perspective, this case highlights several important points. First, recurrent or atypical facial nerve palsy should not be considered idiopathic without thorough investigation. Second, external auditory canal masses with aggressive features and unusual bleeding characteristics should raise suspicion for malignancy. Third, a multidisciplinary approach involving otolaryngologists, radiologists, and hematologists is essential for accurate diagnosis.

CONCLUSION

Recurrent facial nerve palsy associated with atypical external auditory canal lesions should prompt consideration of leukemic relapse, particularly in patients with a history of APL.

Leukemic infiltration may closely mimic cholesteatoma both clinically and radiologically, leading to diagnostic delay. A multidisciplinary approach integrating otologic, radiologic, and hematologic evaluation is essential for accurate diagnosis and timely management.

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