Indian Journal of Otolaryngology and Head & Neck Surgery (2024) 76:3693–3697
https://doi.org/10.1007/s12070-024-04702-y

CLINICAL REPORT

Atypical Presentation of Acute Suppurative Otitis Media with Facial Palsy: Extra Medullary Manifestation of AML in Temporal Bone

Ripu Daman Arora[1] · S B Nidhin[1] · Nitin M Nagarkar[1] · Amit Kumar Banjare[1]

Received: 20 January 2024 / Accepted: 12 April 2024 / Published online: 26 April 2024

Abstract
Acute Myeloid Leukemia (AML) is the most common leukemia observed in the adult population, accounting for 80% of all leukemia cases. Extramedullary involvement in AML, where leukemic cells are found in organs or tissues outside the blood or bone marrow, is a rare occurrence [1]. The most frequent sites of extramedullary disease include the skin, central nervous system (CNS), and lymph nodes [2, 3]. In this case report, we present an instance of extramedullary AML in the temporal bone, which initially presented with symptoms such as earache, discharge, and facial asymmetry, mimicking acute suppurative otitis media with facial palsy. The patient underwent mastoid exploration and facial nerve decompression. A post-operative bone marrow biopsy confirmed the diagnosis of AML, leading to the initiation of chemotherapy. The patient is currently under follow-up care.

Keywords Extra medullary acute myeloid leukemia · Facial palsy · Temporal bone myeloid sarcoma

Introduction

Extramedullary manifestations of acute myeloid leukemia (AML) in the temporal bone are rare but significant. While AML primarily affects the bone marrow, there are instances where leukemic cells infiltrate the temporal bone, leading to various complications such as hearing loss, facial nerve dysfunction, balance problems, headaches, and intracranial pressure issues [1]. Diagnosis involves a combination of clinical evaluation, imaging, and biopsy. Treatment, typically overseen by a multidisciplinary team, may encompass chemotherapy, radiation therapy, and surgical interventions [2, 3]. The prognosis varies depending on the extent of the disease and the patient's response to treatment. The extramedullary manifestation of Acute Myeloid Leukemia poses a unique diagnostic challenge. In this case report, we present an intriguing case of a 29-year-old male patient who initially presented with symptoms resembling acute suppurative otitis media with facial palsy, only to be later diagnosed with temporal bone chloroma.

✉ S B Nidhin
sbnidhin@gmail.com

[1] Department of ENT & HNS, AIIMS Raipur, Raipur, India

Case Report

A 29-year-old male patient presented to our outpatient department (OPD) with complaints of sudden right ear pain and persistent right ear discharge for the past three weeks. The ear pain was described as a sudden, deep, throbbing sensation accompanied by profuse, foul-smelling, mucopurulent discharge that was not blood-stained. Approximately one week later, the patient noticed facial asymmetry. Upon examination, the right ear canal appeared edematous and filled with granulation tissue and pus. A grade 5 facial palsy, according to the House-Brachmann classification, was observed. Figure 1a.

A high-resolution computed tomography (HRCT) of the temporal bone was performed, revealing opacification in the ear canal, middle ear, and aditus without evidence of cavity formation Fig. 2a, b. Notably, the horizontal segment of the facial nerve showed dehiscence Fig. 2c. The initial diagnosis was acute suppurative otitis media with facial nerve palsy, prompting the patient to undergo mastoid exploration and facial nerve decompression.

During the intraoperative examination under a microscope, a total perforation with extensive destruction of soft tissue and necrosis of the external auditory canal (EAC) skin was observed Fig. 4. The mastoid antrum was filled with granulation tissue, and the remaining mastoid air cells

Fig. 1 **(a)** Pre operative picture grade 5 facial palsy 1 **(b, c)** post operative picture, complete closure of eye lids with deviation of lower lip persisiting

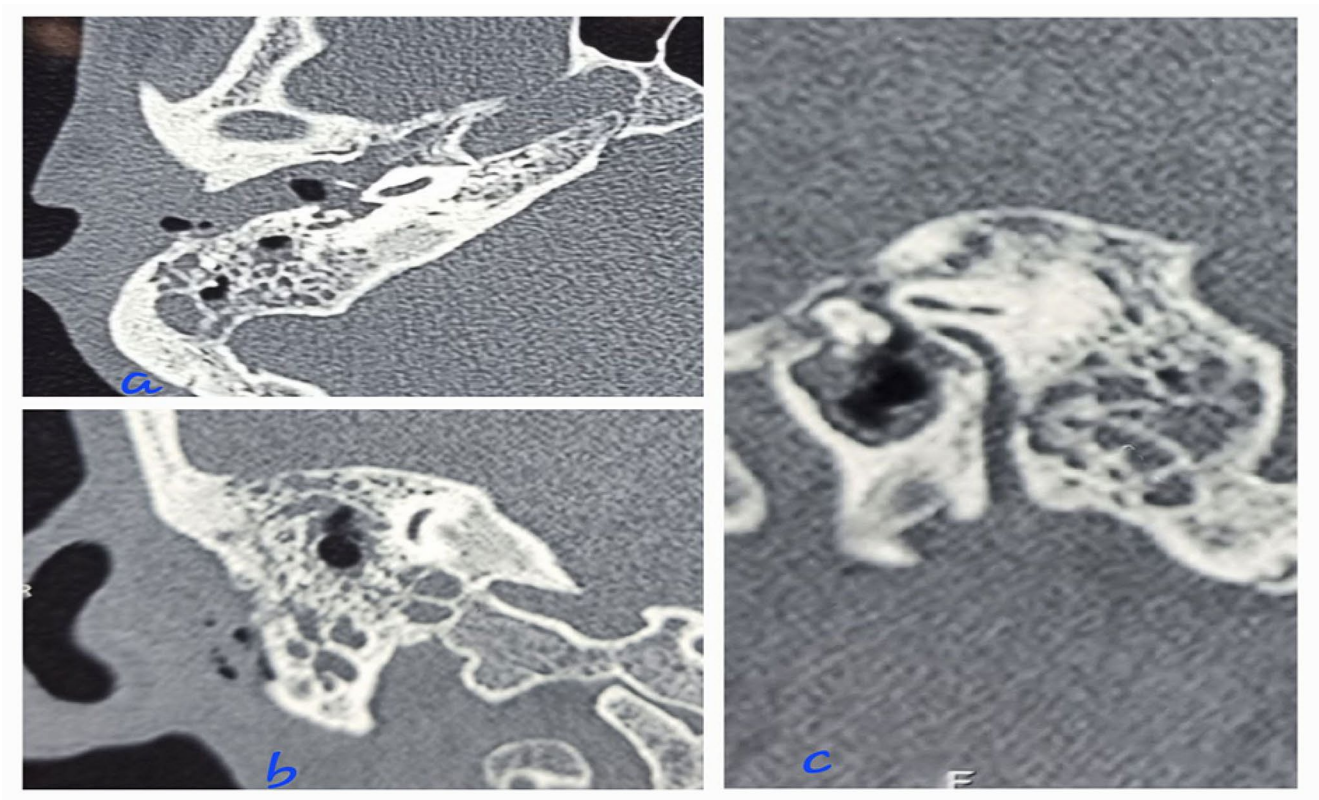

Fig. 2 **(a)** axial cuts; soft tissue filling the EAC and mastoid cavity with bony erosion of EAC **(b)** coronal cuts middle filled with soft tissue, **(c)** saggital cuts demonstration vertical segment of facial nerve canal thinning and erosion

displayed unusually pale bone, a finding uncommonly associated with acute or chronic suppurative otitis media Fig. 3c. Histopathological examination of granulation from EAC and mastoid showed diffusely arranged cells with atypical cells infiltrating fibro-connective tissue Fig. 3b. Post surgery facial nerve palsy improved Fig. 1b, c.

Given the patient's atypical presentation, further investigation revealed a history of multiple blood transfusions for chronic anemia at a local hospital over the past year, which had been initially attributed to hemorrhoids. Subsequent hematological evaluation revealed decreased cell counts across all blood cell lines. A peripheral smear demonstrated pancytopenia. A bone marrow biopsy ultimately

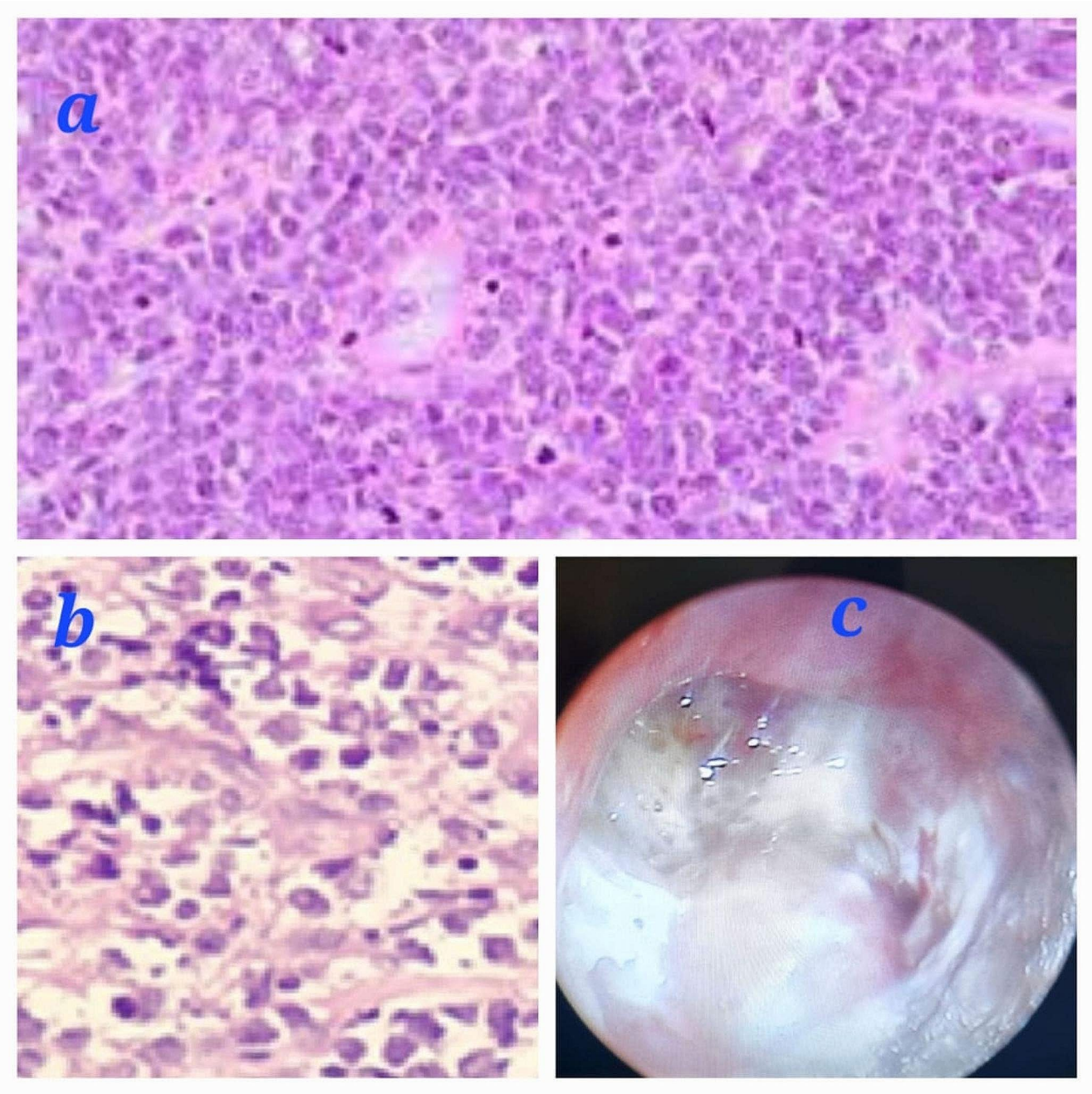

Fig. 3 **(a)** Bone marrow core biopsy H&E section demonstrating clusters of blasts. Blasts exhibit folded nuclear contours with dispersed chromatin. Scattered megakaryocytes are also noted. **(b)** Granulation tissue from EAC and mastoid cavity H&E showed diffusely arranged cells with atypical cells infiltrating fibro-connective tissue. **(c)** Extensive bony ersoion with sclerotic bone exposed and pale granulation in external auditory canal

confirmed the diagnosis of acute myeloid leukemia (AML), further validated by flow cytometry. Figure 3a The patient was promptly initiated on chemotherapy and is currently undergoing treatment.

Discussion

Leukemia is a malignancy marked by abnormal white blood cell proliferation and rarely causes otologic symptoms due to leukocyte infiltration, typically occurring in patients previously diagnosed with leukemia, with children being more commonly affected [4]. In adults, it has been reported as the

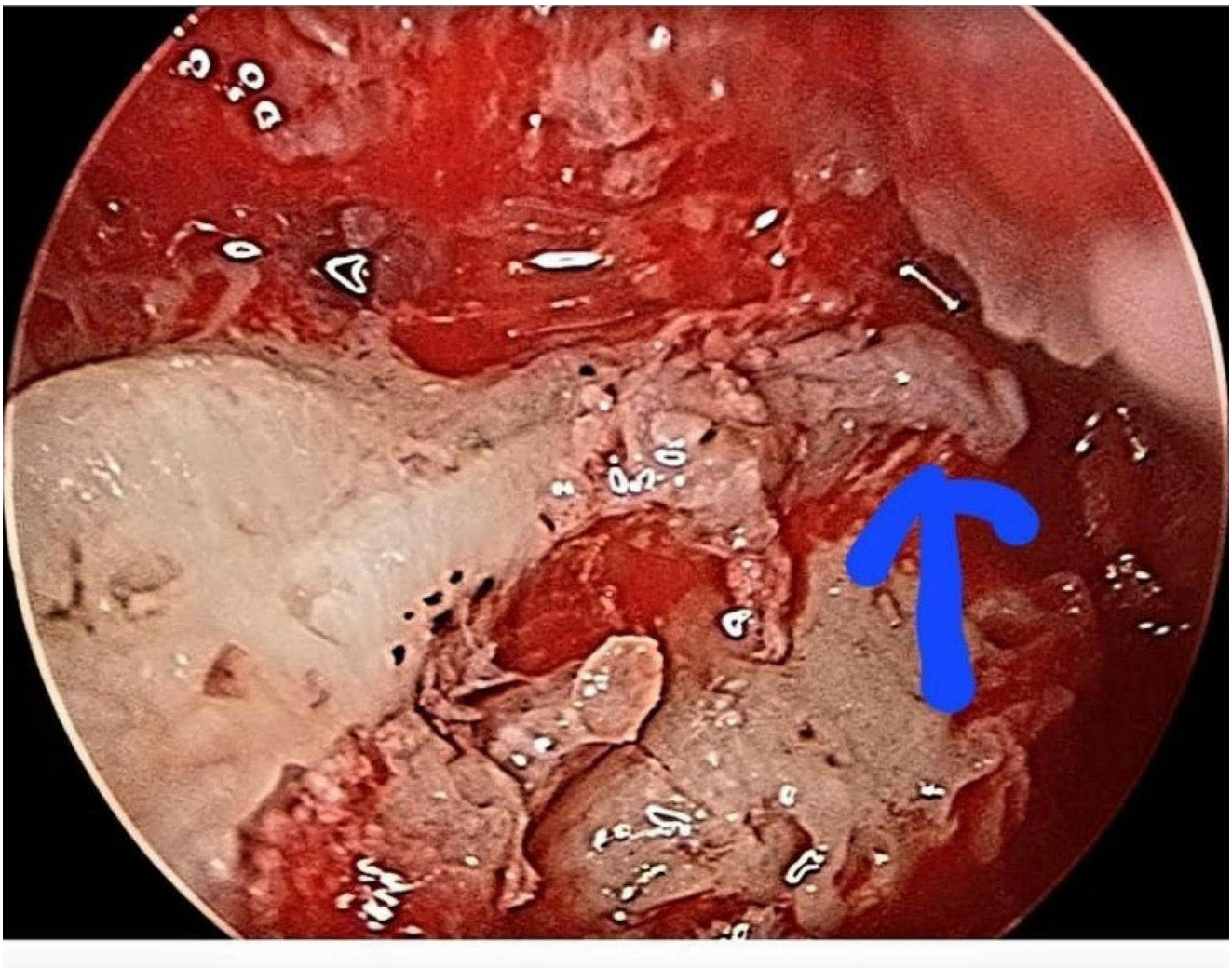

Fig. 4 Intra-operative picture showing dehiscent facial nerve with surrounding granulation tissue

initial sign of AML only once [5]. Granulocytic sarcomas often appear in myeloproliferative disorder patients, sometimes preceding clinical leukemia. These sarcomas typically occur in sites like the periosteum, bone, soft tissue, lymph nodes, and skin. Paperella et al. demonstrated that among 45 temporal bones from 25 leukemic patients, 7 (28%) experienced clinical problems without a clear leukemic cause, while 5 (20%) had otologic complications directly linked to leukemia [6]. Patients with acute mastoiditis typically have a normal mastoid air cell system, unlike those with chronic otitis. Eosinophilic granuloma in the temporal bone typically displays evident bony erosion in radiographic and intraoperative assessments. In contrast, myeloid sarcoma presents with edematous external auditory canal, extensive soft tissue damage, and subtotal perforation with pale granulation. Myeloid sarcoma appears to affect the temporal bone more randomly and diffusely than non-systemic malignancies, as reported by Berlinger [5]. Mastoiditis is more common in cases of leukemic infiltration, whereas petrous apex involvement is more frequently observed in the metastatic spread of non-systemic malignancies.

Our case initially presented as acute mastoiditis, resulting in facial nerve palsy. Consequently, we conducted mastoid exploration surgery. This case prompts questions about the role, timing, and extent of surgery in atypical acute mastoiditis. We believe that surgery was warranted because it reduced the bulk of soft tissue in the middle ear and mastoid cavity, facilitating the collection of a specimen for pathological confirmation. If a tumor, even of an unknown type, is identified, we propose that surgery should be focused on obtaining sufficient tissue for histopathologic diagnosis, reducing tumor size (to enhance the effectiveness of subsequent radiation or chemotherapy), and preserving ear function. If the tumor does not seem to originate from the facial nerve, then decompression of the facial nerve may not be necessary. We concur with Lundberg's recommendation

that "physicians should order leukocyte differential counts as needed on a case-by-case basis." The use of automated WBC differential counts via flow cytometry could have potentially expedited the leukemia diagnosis. However, even if the diagnosis of acute leukemia had been established prior to surgery with suspected granulocytic sarcoma, we believe that surgical exploration of the mastoid, middle ear, and facial nerve decompression to investigate unidentified infection, alleviate edema, and reduce tumor size would have been justifiable, In cases of AML with extramedullary manifestation chemotherapy is the treatment of choice ideally.

In cases where a patient with myeloid sarcoma has leukemia or relapsed leukemia, combination chemotherapy for acute leukemia may induce complete remission. Additional cranio-cephalic radiation therapy is often considered when the disease persists after chemotherapy [7]. High-dose therapies as a frontline approach can better achieve complete remission and possibly cure the disease. Radiotherapy for residual mass and bone marrow transplantation can also improve the outcome of this patient [8].

Conclusion

Extramedullary manifestation of AML in Temporal bone presents as atypical mastoiditis with most common symptom been post aural mass following deafness, vertigo, and facial nerve palsy based on site of deposition.

HRCT temporal bone demonstrates extensive bony erosion of canal and soft tissue deposition in mastoid air cells, intraoperatively extensive soft tissue and bony erosion of canal with granulation tissue in mastoid cells.

Surgery should only be limited to mastoid exploration for getting biopsy and relieving local infection and oedema.

Post operative chemotherapy and loco-regional radiation is the treatment of choice.

Ethical Statement.

This study is in compliance with ethical standards, no conflict of interest, no external funding and prior informed consent is taken from patient and family.

Fundings On behalf of all Co-Authors, I shall bear full responsibility for the submission. I confirm that all authors listed on the title page have contributed significantly to the work, have read the manuscript, attest to the validity and legitimacy of the data and its interpretation, and agree to its submission we also disclose financial or non-financial interests that are directly or indirectly related to the work submitted for publication.

Declaration

Ethics approval and consent to participate This study is in compliance with ethical standards, no conflict of interest, no external funding and prior informed consent is taken from patient and family.

References

1. Vakiti A, Mewawalla P Acute Myeloid Leukemia. [Updated 2021 Aug 17]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. https://www.ncbi.nlm.nih.gov/books/NBK507875/
2. Goh BS, Tang CL, Tan GC (2019) Myeloid sarcoma of temporal bone: a rare manifestation of Relapse Acute myeloid leukemia. Indian J Otolaryngol Head Neck Surg 71:1023–1026. https://doi.org/10.1007/s12070-015-0930-8
3. Yücel L, Başak H, Beton S (2021) Acute mastoiditis with facial paralysis as initial symptom of acute myeloid leukemia relapse: a rare case report. Egypt J Otolaryngol 37:32. https://doi.org/10.1186/s43163-021-00098-6
4. Todd NW Jr, Bowman CA (1984) Acute myelogenous leukemia presenting as atypical mastoiditis with facial paralysis. Int J Pediatr Otorhinolaryngol 7(2):173–177
5. Koutroupas 5BNT, Adams S, Maisel G (1980) Patterns of involvement of the temporal bone in metastatic and systemic malignancy. Laryngoscope 90(4):619–627
6. Gloria-Cruz TI, Schachern PA, Paparella MM, Adams GL, Fulton SE (2000) Metastases to temporal bones from primary nonsystemic malignant neoplasms. Arch Otolaryngol Head Neck Surg 126(2):209–214
7. Paydas S, Zorludemir S, Ergin M (2006) Granulocytic sarcoma: 32 cases and review of the literature. Leuk Lymphoma 47(12):2527–2541
8. Leite da Silveira P, Gonçalves Silva V, RizzatoPaschoal J, Nizam Pfeilsticker L (2015) Bilateral peripheral facial palsy and mastoid infiltration as symptoms of relapsed acute myeloid leukemia. Eur Ann Otorhinolaryngol Head Neck Dis 132(1):41–43