



Case Report of Granulocytic Sarcoma in Bilateral Temporal Bones

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Abstract

Objective: We report an extremely rare case of granulocytic sarcoma in bilateral temporal bones.

Methodology: Case report and a review of related literature.

Results: The patient was initially performed neoplasm ectomy then systemic chemotherapy in his left ears. After surgery the patient got a significant improvement of his left hearing. One year later systemic chemotherapy was established again after a biopsy of his right ear. Nowadays he is still under the treatment of acute myeloid leukemia without recurrence of temporal granulocytic sarcoma.

Conclusion: Granulocytic sarcoma involving temporal bones is rare. Its diagnosis depends on immunohistochemistry. Systemic chemotherapy is the first therapeutic choice. Surgical application should be limited to biopsy and symptomatic relief.

Keywords: Granulocytic sarcoma; Temporal bone; Conductive hearing loss

Introduction

Granulocytic sarcoma (GS) is a localized extramedullary tumor composed of immature myeloid cells. It's an uncommon disease and often described in association with acute myeloid leukemia (AML). In head and neck, it usually involves the skull, gingiva, orbit, facial skin and even paranasal sinus [1-4]. Though rare, GS can affect temporal bone and produce symptoms which are easily confused with other diseases. Here we report a case of GS in bilateral temporal bones.

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Case Presentation

In November 2006, a 36-year-old Chinese man, with a history of AML (subtype M3) which was in remission, presented to our hospital for otalgia and hearing loss in bilateral ears for a month's duration. Previously in December 2000, he was diagnosed as AML (M3) and received chemotherapy; in January 2001 he achieved complete remission, after that he remained in continuous complete remission for 5 years. He denied the history of facial palsy, hearing loss or vertigo.

On admission, otoscopy showed his left external acoustic meatus was completely blocked by a soft-tissue mass. His right external acoustic meatus was stenotic because of swelling of its posterior and anterior portions, only a small part of the right tympanic membrane could be seen, which seemed normal. No pathological enlargement of lymph nodes or organomegaly was noticed. Laboratory studies including complete blood counts and serum biochemistry showed normal values. CT scan of both temporal bones revealed both external acoustic meatus, especially the left, were filled with masses. No bony lesions were found (Figure 1). The CT scan was also suggestive of right mastoiditis (Figure 1). Pure-tone audiogram revealed conductive hearing loss in bilateral ears with air-bone gaps about 35 to 45 dB in the right and 30 to 55 dB in the left (Figure 2).

Because of the patient's history of AML, he was first sent to another hospital to review his leukemia. In that admission, his bone marrow aspirate didn't show any relapse of AML. Though GS couldn't be excluded, other diseases, such as sarcoma, were considered preferentially. Few days later, the patient was submitted to surgery in our hospital. A neoplasm ectomy of the left external acoustic meatus was performed through a retroauricular approach. During the surgery, a tumor was seen originating from the walls of external acoustic meatus, involving part of the tympanic membrane. The tympanic cavity was normal and the ossicular chain was intact. The mass and the tympanic membrane were removed, the ear canal was widened and the temporalis fascia

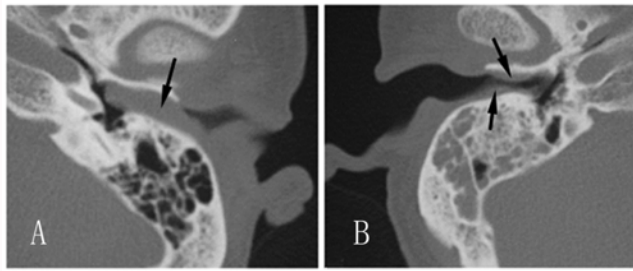


Figure 1: (A) Axial CT scan of the left ear obtained before treatment shows a complete blockage of the external acoustic meatus by a soft tissue mass (arrow), without bone destructions. (B) Axial CT scan of the right ear obtained before treatment shows mass (arrow) in external acoustic meatus causing a stenosis, it also revealed an asymptomatic mastoiditis.

was harvested to rebuild the tympanic membrane. The specimen was performed HE and immunohistochemistry staining. In HE staining, the lesion was consistent showing pleomorphic cells having blastic and hyperchromatic nucleus with large cytoplasm. In immunohistochemistry, it was positive for myeloperoxidase, LCA, lysozyme, CD43, and CD117 (Figure 3), but negative for CD20 and CD7. These results lead to the conclusive diagnosis of GS.

Because of the diagnosis of GS, the patient immediately received a systemic chemotherapy. But due to his poor compliance, only one course of the treatment was finished. One month after the operation, the patient got a complete recovery from his left ear symptoms, and pure-tone audiogram showed a significant increase of air conductive hearing (Figure 2). However, the symptoms of his right ear hardly changed and the otalgia became even severe. In December 2007, the patient received operation of his right ear in our hospital. A lesion similar with the left was seen in the canal, the lesion was completely removed and diagnosed as GS by pathology. After surgery, systemic chemotherapy began. Three months later, the patient recovered from right otalgia, however the hearing hardly improved. Nowadays, the patient is still under the treatment of AML, and clinical examination or CT scans haven't detected any signs of temporal GS recurrence.

Discussion

GS, also termed myeloid sarcoma or “chloroma”, is a rare disease. GS has a close relationship with AML. As reported before, about 2–8 % of patients with AML develop GS [5]. It may present at any time of the course of AML [6]. Though all subtypes of AML could develop GS, the French-American-British (FAB) subtypes M2, M4, M5 are most frequently observed [7]. Certain chromosome abnormalities are

associated with a higher incidence of GS, particularly t (8; 21), and less frequently inv (16) (p13;q22) [8]. GS can also arise in patients with other types of myelo proliferative disorders, such as chronic myelogenous leukemia blastic transformation and myelodysplastic syndrome [9-11].

GS in temporal bone is quite rare. Chapman and Johnson described the first one in 1980 [12]. Temporal GS is most commonly found in patients below the age 30, and its incidence has no deviation in gender or side. GS could span the whole temporal bone, but mastoid is most often involved. Presenting symptoms of temporal GS include facial palsy, otalgia, tinnitus and conductive hearing loss. To our knowledge, GS usually affects unilateral temporal bone; the case presented here is the first one with bilateral temporal GS.

Recognition of GS is important for timely treatment. As long as a patient present for neoplasm with a history of AML, GS should be considered. Some investigations can provide clues for diagnosis. On CT scans, GS may appear as a well-defined enhancing mass sometimes with osteolytic destruction. On MRI, GS may be homogeneously hyper intense on fat saturated T2-weighted images, hypo intense on T1-weighted images and moderate enhancing after intravenous gadolinium contrast administration [13]. However, it's impossible to confirm GS from imaging findings alone. Making a conclusion often needs biopsy for histological diagnosis.

The differential diagnosis of temporal GS comprises external canal cholesteatoma, temporal lymphoma and temporal sarcoma. Temporal GS can be easily parted from external canal cholesteatoma for its lack of keratinized debris. However, it's difficult to distinguish GS from lymphoma and sarcoma by routine investigations. At that condition, immunohistochemical analysis should be performed. The most sensitive markers for GS include CD3, CD43 and myeloperoxidase [14], others such as CD20, LCA and lysozyme can also be used. In addition, the tumor should be negative for B and T cell markers [15].

The optimal treatment of GS is not clear since there is not enough data and large prospective studies in the literature. Nowadays GS is mostly suggested to treat by systemic chemotherapy with or without radiotherapy [16]. Other therapeutic choices include hematopoietic stem cell transplantation and targeted therapy. Surgery alone is not recommended for the high incidence of AML and extramedullary relapse [17]. In this case, we used surgery combining systemic chemotherapy to treat our patient. In our opinion, Because of the anatomic complexity of temporal bone, biopsy in it most frequently needs a surgical approach. Surgery should also be reserved for patients

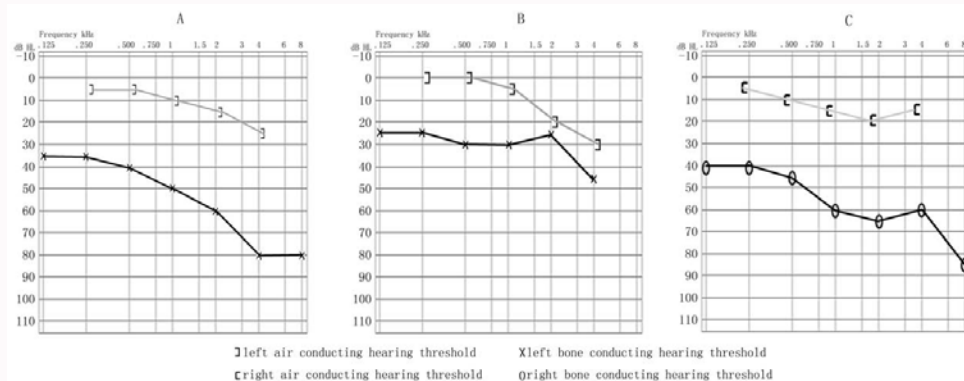


Figure 2: Pure-tone audiogram obtained before the surgery of both ear (A, C) and after the surgery of left ear (B).

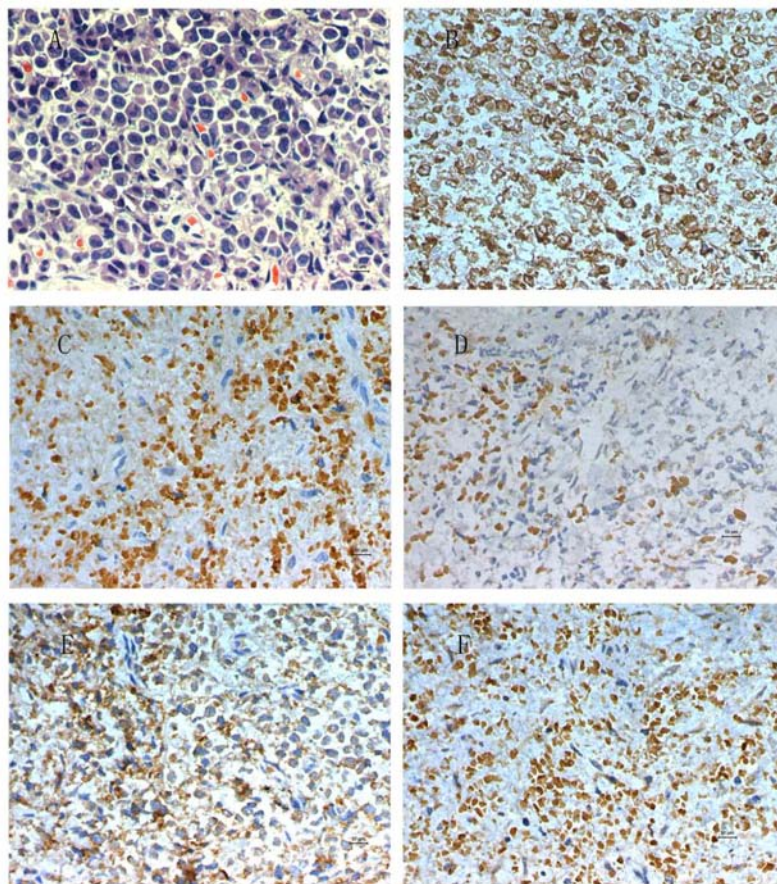


Figure 3: Histology of the surgical specimen from the left ear.

(A) Diffuse infiltration of large mononuclear cells with wide cytoplasm and polymorphic nuclei. Immunohistochemical staining is positive for (B) lysozyme, (C) CD 43 antigen, (D) CD117 antigen, (E) LCA antigen and (F) myeloperoxidase. The bars in photos equal to 10 μ m.

having conductive hearing loss or facial palsy, for its rapid effect of symptomatic relief. To our patient, the surgery not only contributed to biopsy, but also successfully improved his left hearing.

Conclusion

GS involving bilateral temporal bones is extremely rare. It should be considered for patients present for neoplasm with a history of acute myeloid leukemia. Conclusive diagnosis of granulocytic sarcoma needs immunohistochemical analysis. Systemic chemotherapy is the first therapeutic choice. Surgical application should be limited to biopsy and symptomatic relief.

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