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Case report

Facial nerve paralysis in acute otomastoiditis as presenting symptom of FAB M2, T8;21 leukemic relapse. Case report and review of the literature

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Abstract

Granulocytic sarcoma (chloroma) is a rare solid, extramedullary tumour composed of immature granulocytes, occurring during granulocytic leukemia. Leukemic involvement of the temporal bone is not uncommon and may present in a variety of ways. Symptomatic facial nerve paralysis is one of these. The authors report a case of facial nerve paralysis as the presenting symptom of leukemic relapse in a 16-year-old white male, affected by acute myelogenous leukemia FAB M2, karyotype 46xy, T8;21.

Keywords: Acute otomastoiditis; Facial paralysis; Chloroma; Leukemia

1. Introduction

Leukemia is a malignant disease, characterized by abnormal proliferation of white blood cells and their precursors. This hematological disease can affect the external, middle and inner ear, as far as the petrous apex [2,5,19,24,29,36,38,39]. Such involvement depends on: (1) neoplastic infiltration, (2) infections and (3) hemor-

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rhage. Leukemia in the ear can cause bleeding hemorrhages of the external ear and tympanic membrane, skin lesions of the external auditory canal, red or thick tympanic membrane, hemorrhagic or serous acute otitis media with effusion or suffusion of the mucosa of the middle ear, conductive or sensorineural hearing loss, acute otomastoiditis with facial or acoustic cranial nerve paralysis [10,11,16,18,19,20,24,36,38,39].

Although unusual, involvement of the temporal bone may be the presenting sign of leukemia, and a granuloblastic sarcoma or chloroma is the most typical presentation of leukemic infiltration. It is a rare, extramedullary tumour, due to a localized accumulation of immature cells of the granulocytic series, observed in 3.1–6.8% of granulocytic leukemia [17,20,22].

Granuloblastic sarcoma was first described in the lacrimal gland by Burns in 1811 [3]. The term chloroma was first introduced in 1853 by King [13], because of its greenish colour, as a result of myeloperoxidase activity in the tumour cells. Dock in 1893 [7] recognized the association with leukemia. Granuloblastic sarcoma usually arises in patients with overt leukemia; however it may occur during clinical and hematological remission [26]. Sometimes it can even represent the first symptom of leukemia [14], months or years before the diagnosis of 'acute myelogenous leukemia' is made — as much as 26 months earlier [32]. The initial solitary lesion may be misdiagnosed as lymphoma or eosinophilic granuloma [21,37].

2. Case report

R.P. was a 16-year-old white male, admitted to hospital with signs and symptoms of endocranic hypertension. At the time of admission the patient showed anemia (Hgb, 8.4 g/dl), thrombocytopenia (42×10^9/l) and hyperleukocytosis (WBC, 15.6×10^9/l) with 75% of immature cells at differentiated blood-cells count. Bone marrow aspiration confirmed the diagnosis of acute leukemia FAB M2. No extramedullary disease was present at that time and cranial CT-scan did not show central nervous system involvement. Cerebro-spinal fluid was not examined due to the poor clinical status of the patient. He received standard chemotherapy (daunorubicin and cytosine arabinoside) and he obtained a complete hematological remission in August 1987. No HLA-identical donor was found in the family and the patient was submitted in February 1988 to autologous unpurged bone marrow transplantation (conditioning regimen: busulfan and cytoxin). In January 1989, the patient showed a leukemic relapse and was treated with chemotherapy including idarubicin and cytosine arabinoside, obtaining a second complete hematological remission, in March 1989. Despite maintenance treatment, in May 1990 the patient presented a second medullary relapse together with a peripheral paralysis of the seventh cranial nerve, otalgia, tinnitus and severe conductive hearing loss. A new examination of the cerebro-spinal fluid did not reveal leukemic infiltration.

At otologic examination, the patient showed an edematous infiltration of the left mastoid with disappearance of the post-auricular groove; the external auricular canal was very stenotic because of the lowering of its postero-superior portion.

Only the antero-inferior portion of the tympanic membrane was visible and it appeared hyperemic and swollen without otorrhea. A CT-scan of the head showed opacification and sclerosis of the mastoid air cells, with granulation tissue occluding the middle ear; no bony lesions were found, not even in the ossicular chain (Fig. 1). The clinical and radiological findings were suggestive of acute left otomastoiditis, together with peripheral omolateral paralysis of the seventh cranial nerve.

The patient was submitted to surgery and an infiltration of post-auricular soft tissues by a gelatinous mass protruding into the external auricular canal and totally taking up the tympanic cavity was observed. After the exploration of the mastoid up to the petrous apex, the tegmen antri appeared worn with the

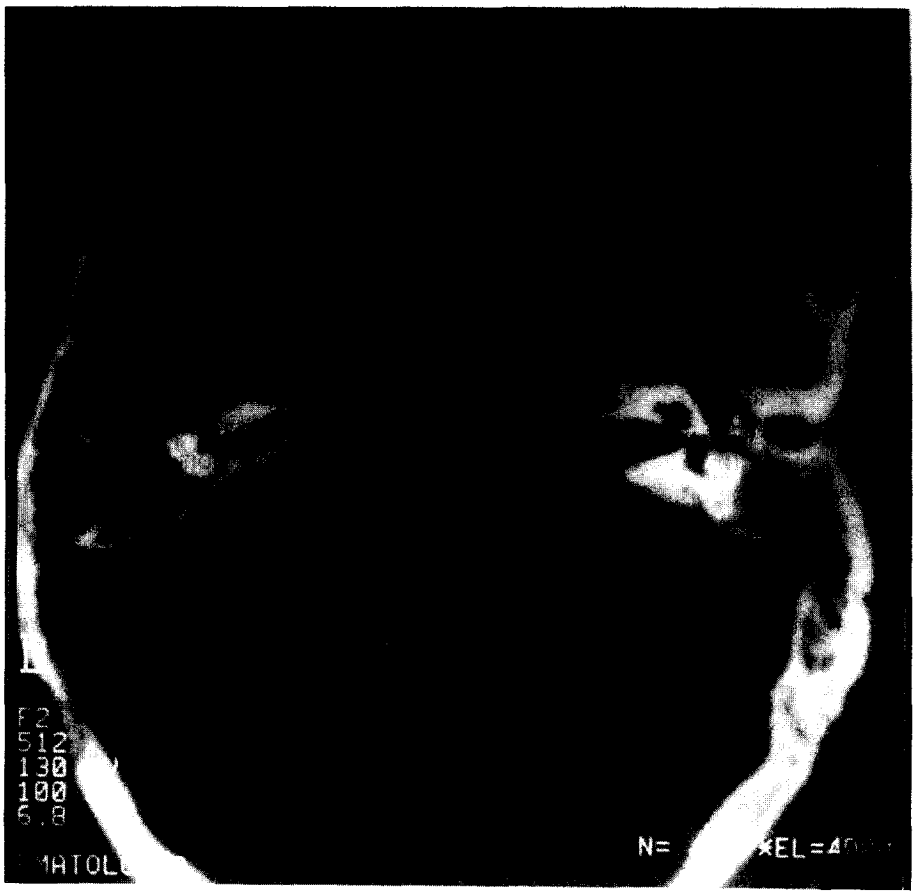

Fig. 1. The CT scan of the temporal bone shows the considerable opacification of the left mastoid air cells, with a soft tissue density in the middle ear cavity. No bony erosion is present and the ossicular chain is complete.

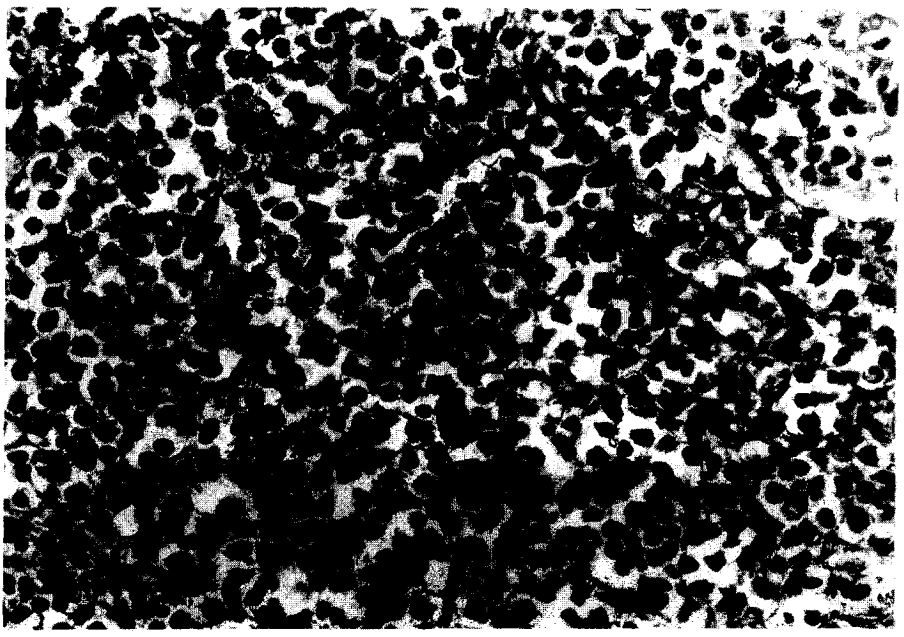

Fig. 2. Histologic section demonstrating the sheets of blastic cells, with cytoplasm granulated, consistent with a diagnosis of granuloblastic sarcoma; magnification: × 25, H and E.

procidentia of a congestive meninx. After removing the hyperplastic granulations, the gelatinous mass and the ossicular chain except for the stapes, the seventh cranial nerve appeared slotted and uncovered, between its second and third portion. Subsequent to the unbridling of the bony canal and the decompression of the nerve, the last revision of the surgical cavity up to intact bone was performed. The packing of the bony cavity was performed last, together with a reconstruction of the external acoustic canal. Histological examination showed a blast cell infiltration, some of which granulated, suggestive of extramedullary leukemia (Fig. 2).

The post-operative course of the patient was uneventful with a partial resolution of the facial nerve paralysis. He received a new cycle of chemotherapy (idarubicin, etoposide and cytosine arabinoside) obtaining a third complete hematological remission. Two months after surgery, clinical examination and radiological imaging showed a new leukemic infiltration in the large residual surgical cavity (Fig. 3). In July 1990, the patient developed pneumonia caused by *Acinetobacter anitratus*, during hematological remission. The clinical status worsened until October 1990, when acute neurological symptoms appeared, including diplopia and nystagmus. Despite no significant alterations of the cerebro-spinal fluid and cranial CT-scan, the neurological signs rapidly worsened and after the onset of an acute inflammatory polineuropathy the patient developed respiratory failure and coma. He died in the intensive care unit in June 1991.

3. Discussion

The involvement of temporal bone during leukemia has been reported since the beginning of this century [19] on post-mortem examinations; the first clinical case go back to 1950s (Table 1).

The incidence of granulocytic sarcoma is similar in acute and chronic granulocytic leukemia [17]; and fourfold higher in myelogenous leukemia FAB M2, characterized by typical chromosomic translocation T 8;21, than in the general leukemic population [34,35]. The development of granulocytic sarcoma in patients with leukemia implies a poor prognosis, often heralding a blastic crisis [23]. There appears to be no sex predominance, but 40% of cases are described in patients under 15 years of age [17]. Chloroma can arise at any site but it is most commonly found in bone (temporal bone in 50% of cases, [25]) and nervous tissue (particularly the orbit and the epidural space) and can be absolutely asymptomatic [15,17]. The

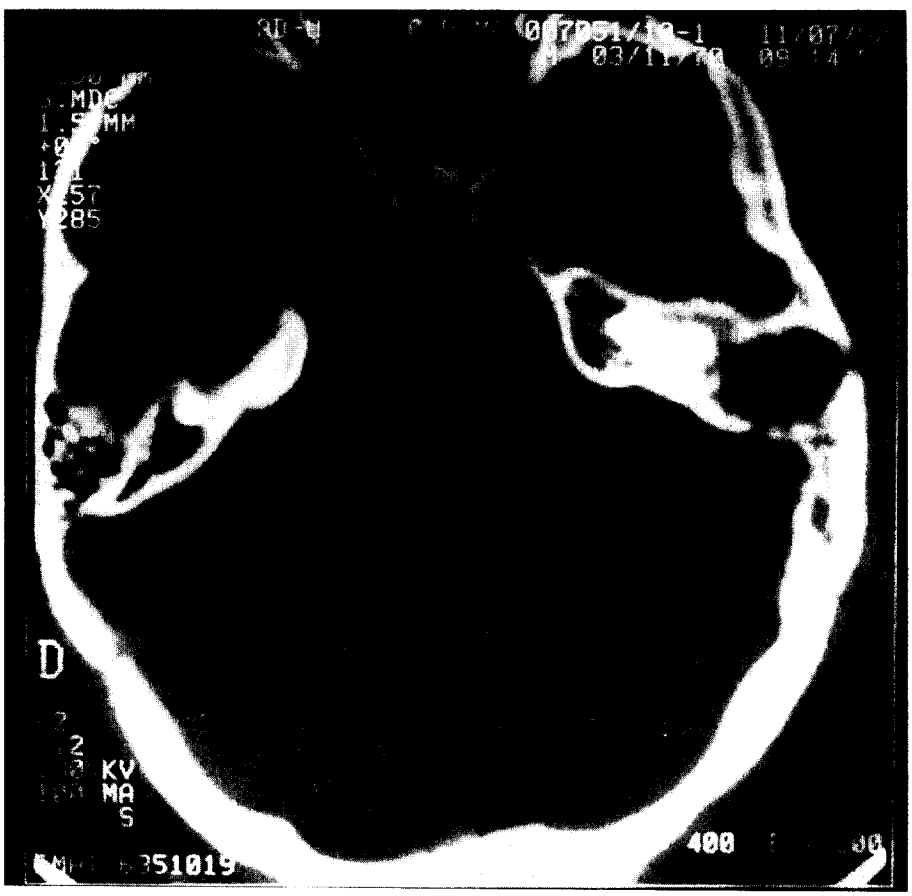

Fig. 3. A post-operative CT scan of the temporal bone which shows the relapse of granuloblastic sarcoma in the wide residual middle ear cavity (2 months after surgical intervention).

Table 1
Review of the literature

Reference	Leukemia	Cases	Site	Facial paralysis
Manasse [19]	AML	1[a]	Inner ear	-
Druss [8]	ALL	1	Middle ear	+
Abba and Manoccini [1]	AML	1	Middle ear	+
Mari et al. [20]	AML	1	Middle ear	-
Bustamante-Balcarcela and Savin-Vazquez [4]	ALL	1	Middle ear	-
Zechner and Altmann [40]	LLC	6	Middle ear	-
	ALL	5		
	AML	2		
	SCL	8[b]		
Wright [38]	AML	1	Middle ear	+
Paparella et al. [24]	ALL	4	Middle ear	-
	AML	1		
Sklansky [33]	ANLL	10	Middle ear	+
	ALL	14		
Chapman and Johnson [5]	AML	1	Middle ear	+
Todd and Bowman [36]	AML	1	Middle ear	+
Zappia et al. [39]	ANLL	1	Middle ear	+

[a]Post-mortem examination.
[b]Stem cell leukemia.

most common presenting symptom in temporal bone involvement is an aching post-auricular swelling, associated with a generally conductive hearing loss, otalgia and tinnitus, until the onset of an acute otomastoiditis, caused by bacterial superinfection [5,16,28,36]. Eustachian tube obstruction or infiltration, secondary to leukemic involvement of the nasopharynx, suffusion and/or effusion of the middle ear mucosa, can lead to acute otomastoiditis [31]. Massive bony erosion, deafness and vertigo may develop at a later stage due to progression of the lesion, if untreated [8,9,11,30,36].

Acute otomastoiditis subsequent to leukemic infiltration of the temporal bone may cause facial and acoustic nerve paralysis. The mechanism in this case is the perineural and/or meningeal leukemic infiltration associated with hemorrhage, edema or adverse reaction to drugs [6,12,20,27].

Generally, a wide myringotomy shows that the middle ear cavity is filled with a gelatinous fluid; mastoidectomy shows that the hypotympanum and the epitympanum too are filled with a greenish soft tissue that overlies the antrum and the second genu (horizontal portion) of the seventh cranial nerve. Mostly there is no evidence of bony dehiscence along the Fallopian canal. Leukemic infiltration can engulf the ossicular chain. Such anatomical findings are in agreement with the radiological aspects. CT-scan of the head shows opacification without trabeculation of the mastoid air cells, and middle ear cleft with considerable swelling of the mucosa. That is consistent with a leukemic focus of chloroma.

The surgical management of chloroma is restricted to obtaining a tissue diagnosis, draining infection and reducing neoplastic infiltration before chemotherapy and/or radiotherapy. If the tumour does not seem to originate from the facial nerve, its decompression would not seem appropriate [36]. If the leukemic involvement is localized in the petrous bone, local low-dose radiotherapy and systemic chemotherapy are the treatments of choice. Finally, if leukemic cells have spread into the cerebrospinal fluid, either by direct extension or through the cochlear duct, intrathecal chemotherapy with cytosine arabinoside should also be considered.

In patients affected by granulocytic sarcoma, a leukemic metastasis of the temporal bone should be considered, particularly when peripheral facial paralysis is present. CT scan of the head after contrast mean injection is mandatory to rule out neoplastic infiltration. In fact, in the case reported above, we thought it unnecessary to perform a CT scan with contrast, being the diagnosis of classic otomastoiditis with peripheral facial paralysis apparently obvious.

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