CASE STUDY

Temporal Bone Myeloid Sarcoma
Sarcoma mieloide del hueso temporal

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Received 21 March 2013; accepted 20 September 2013

Case Report

A 69-year-old male previously diagnosed with acute myeloblastic leukemia secondary to myelodysplastic syndrome (LMA with dysplasia, karyotype 46XY, and no cytogenetics abnormalities) refractory to treatment (hydroxyurea, cytarabine, mitoxantrone, and azacitidine) presented with right-side otalgia, otorrhea, and deafness with 2 months progression. He had been treated with oral ciprofloxacin, eardrops with dexamethasone + neomycin + polymyxin B and ibuprofen without success. Otoscopy showed polypoid tissue in the concha of the auricle, otorrhea, and tympanic membrane purple bulging (Fig. 1). Patient was hospitalized with the suspicion of malignant external otitis, and intravenous ciprofloxacin, ceftazidime, and meropenem were prescribed. Investigations revealed hemoglobin 8.6 g/dL, leucocytes $43.2 \times 10^3$ g/L, platelets of $24 \times 10^3$ g/L, and negative blood cultures. Head computerized tomography (CT) demonstrated tissue fullness of right antrum, mastoid, and tympanic cavity, with extension to external auditory canal and infiltration of periauricular tissue and petrous bone erosion (Fig. 2A). A biopsy of the concha of the auricle was done and confirmed leukemic infiltration (Fig. 2B). Patient developed mental confusion and right motor lateralization and a new CT was performed. The CT showed voluminous expansive lesion of the right ear, lesions in both cerebral

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hemispheres compatible with chloromas. With these results the diagnosis of myeloid sarcoma of temporal bone was done and the patient started on chemotherapy (cytarabine and hydroxyurea). Patient was transferred to the hospital of his area of residence for palliative care. Follow up was lost, because he was called by letter and did not come.

Discussion

Myeloid sarcoma, also known as chloroma is a rare extramedullary collection of immature cells with myogenous differentiation.\(^1\)\(^-\)\(^4\) It occurs in 3%–8% of patients with acute myeloid leukemia.\(^7\) Although it can occur anywhere throughout the body, it is more common in bone, periosteum, soft tissue, lymph nodes, and skin.\(^1\)\(^-\)\(^2\)\(^-\)\(^4\) In the head and neck, it has been reported to occur in the maxilla, soft palate, nasopharynx, lip, salivary glands, mandible, and temporal bone.\(^1\) Although leukemic infiltration into the middle and inner ears was found to occur in 20% of patients with leukemia, frank myeloid sarcoma of the temporal bone is rare.\(^1\)\(^-\)\(^3\) It may develop de novo or concurrently with the leukemia or after remission, with myeloproliferative disorder or myelodysplastic syndrome.\(^1\)\(^-\)\(^3\)

The single most important point in the diagnosis of temporal bone malignancy is suspicion.\(^5\) Maddox emphasized the symptom triad of otalgia, periauricular swelling, and facial nerve paresis as being highly suggestive of malignant involvement of the temporal bone.\(^3\) The symptoms may mimic infectious pathology.\(^9\)

Biopsy is the best diagnostic method but it is not always possible. A biopsy sample may be difficult to obtain, especially when the mass is in an anatomically difficult site such as the petrous bone. Even if the mass is located in a site that is easily reached, biopsy is not feasible for patient with thrombocytopenia or acquired consumption coagulopathy.\(^5\)

The pathologic diagnosis of myeloid sarcoma can be achieved by positive results of either naphthol ASD chloroacetate esterase (NCAE), immunoperoxidase staining, lysozyme or CD34 antigen to confirm transformation to acute myelogenous leukemia.\(^1\) The term chloroma reflects its greenish hue secondary to the presence of intracellular myeloperoxidase.\(^1\)\(^-\)\(^3\)\(^-\)\(^6\)

Both CT scan and MRI are diagnostically very useful.\(^5\) On CT scans, we can find a well defined area of increased attenuation with a peripheral zone of enhancement.\(^1\) When the bone is affected, lesions appear lytic rather than sclerotic.\(^1\)\(^-\)\(^3\) On MRI images it shows isointense signal intensity to that of muscle or marrow on T1 and T2 weighted images and enhances markedly and homogenously post gadolinium.\(^4\) These lesions may mimic lymphoma, pseudotumor or meningioma on imaging studies.\(^1\)\(^-\)\(^3\) Pathological confirmation is important and in most cases the final diagnostic clue.\(^3\)

There is no standard treatment for myeloid sarcoma, especially when occurs de novo, rather the treatment is similar to that of systemic disease, as it is considered a manifestation of such.\(^3\) When a patient with myeloid sarcoma has leukemia or relapsed leukemia, combination chemotherapy for acute leukemia may induce complete remission.\(^6\) Additional craniocephalic radiation therapy is often considered when the disease persists after chemotherapy.\(^2\) Recent literature suggests that early, aggressive induction chemotherapy may prolong survival or lead to complete remission.\(^1\) The prognostic of myeloid sarcoma is unfavorable.\(^5\) The mean survival time has been reported to be from 2.5 months to 22 months.\(^1\)

We report an unusual case of temporal bone myeloid sarcoma. Recognition of this rare entity is important, because early aggressive chemotherapy can cause regression of the tumor and improve prognosis.

Conflict of Interest

The author declares no conflict of interest.

References