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Myeloid sarcoma of submandibular salivary gland

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Abstract

Objective: To report a rare case of a myeloid sarcoma of submandibular salivary gland.

Methods: A 65-year-old woman with a history of successfully treated myelodysplastic syndrome, presenting with periodic painful swelling of her right submandibular area.

Results: Physical evaluation, ultrasound and CT scan revealed the presence of a 3-cm mass contiguous to the submandibular salivary gland. A core needle biopsy confirmed the diagnosis of myeloid sarcoma. Bone marrow biopsy was still showing complete remission and the submandibular gland was the only extramedullary site involved. The patient was submitted to chemotherapy.

Conclusion: Myeloid sarcoma is a rare extramedullary neoplasm. It can virtually involve any anatomic site, but it usually involves lymph nodes, paranasal sinuses, skin, soft tissue and periostium. Myeloid sarcomas of salivary glands are very rare and ENTs should be aware of this disease in order to include it in the differential diagnosis of a solitary neck mass.

Keywords

Myeloid sarcoma, extramedullary neoplasm, submandibular gland

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Introduction

Myeloid sarcoma (MS) is a rare extramedullary neoplasm composed of immature or mature granulocytes or monocytes. In the literature, it is known by a variety of names including granulocytic sarcoma, monocytic sarcoma, myeloblastoma and chloroma. In 2%–8% of cases MS occurs before, during or after the onset of acute myeloid leukemia (AML) and, more rarely, in patients with myeloproliferative disorder. It is extremely rare its presentation without any prior history of myeloid neoplasm.[1–3]

It has been reported an increase in MS following allogenic stem-cell transplantation and some authors have suggested that it may represent a reduced graft-versus-leukemia effect at extramedullary sites.[4]

MS can involve any anatomic site; however, lymph nodes, periostium, paranasal sinuses, soft tissue and skin are most commonly affected. The involvement of salivary glands is very rare and there are just a few cases reported in the literature.[2,5,6]

Case report

In January 2015, a 65-year-old woman presented with right submandibular slowly enlarging mass. Patient's past history included a myelodysplastic syndrome diagnosed in 2010. In 2011, after induction and consolidation chemotherapy, the patient had undergone allogenic hematopoietic stem-cell transplantation with a smooth, event-free course until July 2014. From that time on she had started with periodic painful swelling of the right submandibular area without strict correlation with eating. At that time patient underwent ultrasound, which showed a slightly enlarged right submandibular salivary gland with signs of chronic inflammation. Blood tests were normal and bone marrow examination did not show any signs of relapse. Chronic sialoadenitis was suspected, and whenever the problem recurred, she was treated with oral antibiotics and steroids with full, but temporary, recovery. After 4 months and three acute relapsing episodes, she underwent again echotomography showing a growing salivary mass with some calcifications inside. A CT scan without contrast was prescribed confirming the presence of a mass contiguous to the salivary gland of $2.5 \times 3\ cm^2$.

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Physical evaluation revealed a firm, non-tender, mobile, 3-cm mass, palpable through the floor of the mouth. ENT examination was otherwise normal.

Blood tests were normal and fine-needle aspiration biopsy (FNAB) was performed. Since the findings, immediately evaluated by the pathologist, were not diagnostic, a core needle biopsy (CNB) was performed. Histopathologic examination demonstrated a diffuse and dense infiltrate of hemato-lymphoid cells within the salivary gland parenchyma. These atypical cells were medium sized and had mildly basophilic cytoplasm and round nuclei, with fine chromatin and inconspicuous nucleoli. Immunohistochemical examination was diagnostic for immature myeloid-monocytes expressing myeloperoxidase, CD68 (clone PGM-1) and CD34. The conclusion was MS with French–American–British M4.

Bone marrow biopsy was still showing complete remission and the submandibular gland was the only extramedullary site involved, as a total-body CT scan showed. The patient was submitted to chemotherapy.

Discussion

MS is a rare neoplasm, which can very infrequently affect salivary glands. In the literature, there are very few reported cases of MS involving salivary glands and just a part of those involved the submandibular gland.

The rarity of MS makes this kind of diagnosis challenging and it can be missed or delayed if the possibility of MS is not included in the differential diagnosis. From this point of view, the role of ENT is crucial, being one of the first physicians asked for a consultation in such cases. From a diagnostic point of view, excision of the gland is often not needed, although FNAB is insufficient and CNB is usually required. Because of its rarity, MS can be misdiagnosed, the most common alternative diagnoses being lymphoma, undifferentiated cancer, malignant melanoma, extramedullary hematopoiesis and inflammation. For this reason, careful pathologic evaluation including immunohistochemistry is crucial for its accurate diagnosis.[2,3]

Therapy usually does not include surgery because MS has to be intended as a local manifestation of a systemic disease.

Declaration of conflicting interests

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Informed consent

Verbal informed consent was obtained from the patient for her anonymized information to be published in this article.

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