CASE REPORT

Myeloid sarcoma of the maxillary bone

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Myeloid sarcoma (MS) is a malignant tumour of myeloblasts rarely occurring in the maxillary bone. The tumour may precede or be concurrent with leukaemic infiltration of the bone marrow or herald blastic transformation of a myelodysplastic syndrome or a chronic myeloproliferative disorder. Myeloid sarcoma is uncommon in the oral cavity, but it can involve the palate, gingiva, extraction socket, and cheek. Recognition and diagnosis of myeloid sarcoma involving the soft tissues of the oral cavity in an otherwise asymptomatic patient is important and mandates an appropriate haematological diagnostic workup.

We herein report on a new case without any evidence of haematological disorders. We discuss the pathological diagnosis and the therapeutical approaches.

Case report

An 84-year-old edentulous woman presented with an ulcerated, nodular, infiltrative mass on the right hard palate of recent onset (Fig. 1). Previous medical and dental clinical history was unremarkable. The lesion was biopsied because it was considered clinically suspicious for carcinoma.

The surgical specimen consisted of a 2.0 cm fragment of ulcerated oral mucosa that was entirely occupied by a soft white nodule on cut section. Histologic sections demonstrated extensive infiltration of the submucosa by sheets of small to medium size blastic-like cells that exhibited a high degree of mitotic activity (Fig. 2). All the surgical margins were involved. At higher magnification the tumour cells exhibited a high nuclear/cytoplasmic ratio; nuclei were round to ovoid and frequently appeared indented or “bean”-shaped. The nuclear chromatin pattern was very fine; nucleoli were small and inconspicuous. Tumour cell cytoplasms were slightly basophilic on Giemsa stain, and lacked cytoplasm granules.

The composite findings were suspicious for a high-grade malignant lymphoma. Immunophenotyping was performed with the following monoclonal antibodies: anti-CD3, anti-CD20/L26, anti-CD43/MT1, anti-CD45/Pan Leu and anti-CD45RO/UCHL1. Anti-CD34, TdT and anti-CD10/Calla monoclonal antibodies were also included because the blastic appearance of cells was more suggestive of a precursor cell than a peripheral cell high-grade lymphoma. Markers for myeloid/monocytic differentiation (myeloperoxidase and CD68/KP1) were also tested together with staining for chloroacetate esterase. Proliferation activity was assessed with Mib-1 antibody.

Immunohistochemistry revealed that neoplastic cells were positive for CD45, CD43 and CD34 and that 90% of the cells were Mib-1 positive (Fig. 3). Markers for myeloid/monocytic differentiation (myeloperoxidase, CD68/KP1) were found to be diffusely expressed by cells (Table 1). Chloroacetatoesterase was found to be negative. The CD3 and CD20 were negative.

The diagnostic impression was granulocytic variant of myeloid sarcoma (Fig. 4). A cranial CT scan revealed that the lesion extended from the maxillary alveolar bone with infiltration into the maxillary sinus mucosa. The patient was subsequently referred for a hematological work-up to exclude other synchronous myeloproliferative disorders. No alterations of the peripheral blood profile were noted. A bone marrow trephine biopsy showed normocellular marrow with regular maturation of the three precursor cell series. CD34 was also tested on bone marrow sections to evaluate the percentage of blastic precursor cells. The CD34 positive cells were rare (less than 1%) and were thus interpreted as within the normal range for hematopoietic precursors.

On the basis of the composite clinico-pathological data, the maxillary tumour was finally interpreted as an isolated focus of myeloid sarcoma, granulocytic variant, and was surgically removed (Fig. 5) with right
hemimaxillectomy. The surgical specimen showed a neoplasm that diffusely infiltrated the mucosa of the hard palate; it extended into the maxillary sinus, which was completed filled with neoplastic elements. Tumour was present at all surgical margins.

The patient underwent radiotherapy to the involved field. She has been followed for 7 months. To date, she is alive, well and free of disease.

Comments

Myeloid sarcoma is a localized, solid, extramedullary tumour consisting of immature myeloid precursor cells (1). Burns (2) reported the lesion as long ago as 1811 and termed it chloroma because the tumour often exhibited a greenish colour, resulting from the presence of myeloperoxidase (verdoperoxidase) in the tumour cells. The tumour occurs in association with malignant haematopoietic diseases involving the myeloid series. Such conditions include acute nonlymphoblastic leukaemia, various myeloproliferative syndromes such as chronic myeloproliferative syndromes and, in particular, chronic myeloid leukaemia in blast crisis (3, 4). Therefore, a clinical diagnostic suspicion of myeloid sarcoma must be considered whenever an extramedullary mass appears in a patient with a hematological disorder. However, microscopic diagnosis can be challenging in cases lacking a history of any known hematologic abnormality, since MS can mimic malignant lymphoma or an undifferentiated carcinoma.

Myeloid sarcoma is uncommon in the oral cavity, but it can involve the palate, gingiva, extraction socket, and cheek (5, 6). Isolated myeloid sarcoma of the maxillary bone has been rarely reported. The differential diagnoses for an ulcerative oral mucosa lesion include traumatic ulcer, mucosa ulceration secondary to underlying systemic disease, and primary or metastatic cancer. In this case, the initial clinical impression was carcinoma of the hard palate. Only biopsy yielded to the correct interpretation of the lesion. Immunohistochemistry was very useful because the tumour cells showed no evidence of myeloid differentiation, like cytoplasmic azurophilic granulations on Giemsa stain or histochemical staining with chloroacetateesterase. We can postulate that fixation in formalin could interfere with chloroacetate esterase expression. When tumour cells are poorly differentiated, as they were in this case, granulocytic sarcoma can be mistaken for a malignant lymphoma.
most particularly of lymphoblastic, Burkitt and large cell type. In our case, positive staining with CD43 but not CD3 was an important clue pointing to a myeloid neoplasm. Reactivity for CD34, myeloperoxidase and CD68 enabled us to avoid a diagnostic misinterpretation. In a case like our patient’s, distinction of one subtype from the other variants of MS is difficult on the basis of histomorphology alone. The coexpression of these two markers (myeloperoxidase and CD68/KP1) was helpful and supported granulocytic differentiation of the tumour.

Recognition and diagnosis of myeloid sarcoma involving the soft tissues of the oral cavity in an otherwise asymptomatic patient is important and mandates an appropriate hematologic diagnostic workup. Its diagnosis has a clinical relevance because it can be the first manifestation of an acute non-lymphoid leukaemia. Further clinical haematological investigations, including a bone marrow biopsy, are therefore recommended in each case of myeloid sarcoma. In some cases, the extramedullary tumour can represent a blastic transformation of a myelodysplastic syndrome or a chronic myeloproliferative disorder and there can be a less severe lesion in the bone marrow. The occurrence of myeloid sarcoma therefore carries prognostic significance, because it may signify a blast crisis and a poor prognosis. It is recommended for all cases of apparently isolated myeloid sarcoma to be treated with an aggressive approach, with the specific regimes similar to those used in cases of acute myeloid leukaemia.

References


Table 1 Immunohistochemical results for the differential diagnosis

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<th>Immunoreactivity</th>
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<td>CD68/KP1</td>
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Figure 4 Cytoplasmic myeloperoxidase immunoreactivity supported the diagnosis of a myeloid tumour.

Figure 5 Myeloid sarcoma specimen after surgical excision.