CASE REPORT

Myeloid sarcoma: Case report with an unusual presentation in radicular cyst capsule

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Summary Myeloid sarcoma is a tumour mass of myeloblasts or immature myeloid cells occurring in an extra-medullary site or in a bone. The tumour may precede or occur concurrently with acute or chronic myeloid leukaemia or with other types of myeloproliferative disorders or myelodysplastic syndromes. Granulocytic sarcoma is a rare presentation in the oral and maxillofacial region, but has been reported in oral soft tissues as well as in intra-osseous sites. The majority of these cases occur in patients with known leukaemia or those who eventually develop the disease. Our report details the presentation of myeloid sarcoma in radicular cyst capsule, being also one of the unusual intra-osseous presentations of this rare tumour.

KEYWORDS Myeloid sarcoma; Granulocytic sarcoma; Radicular cyst; Cyst capsule

Introduction

Myeloid sarcoma (granulocytic sarcoma, chloroma) is a tumour mass of myeloblasts or immature myeloid cells occurring in an extra-medullary site or in a bone. The tumour may precede or occur concurrently with acute or chronic myeloid leukaemia or with other types of myeloproliferative disorders or myelodysplastic syndromes.1,2 The incidence of myeloid sarcoma in the course of acute myeloid leukaemia has been reported to range from 3% to 4.7%. The occurrence of extra-medullary lesions before the onset of overt disease is rare.3 The most common sites of occurrence are subperiostal bone structures, lymph nodes and skin.4 Myeloid sarcoma occurs at any age; however, two thirds of cases occur in young people, usually before the age of 15. Oral involvement is rare, but has been reported in oral soft tissues as well as in intra-osseous sites.4–9 The majority of these cases occur in patients with known leukaemia or those who eventually develop the disease.3,4,10

Report of case

A 59-year old man with a fresh diagnosis of acute myeloid leukaemia was referred to oral examination and panoramic tomography because of poor dental status. In the peripheral
blood sample half of the leucocytes were blasts, the bone marrow sample was diagnosed to be hyperplastic and 90% of nuclear cells of the bone marrow were blast cells (Fig. 1). Intra-orally the maxillary and mandibular gingiva were covered by a thick hyperplastic tissue harbouring decayed residual teeth (Fig. 2). Radiographic examination revealed a diffusely bordered radiolucency in the apical area of residual roots of right maxillary canine and lateral incisor. Radioluencies were further found in the marginal border of left maxilla and in the mandible, all related to residual roots (Fig. 3). The lesions were surgically explored and the roots removed. The biopsy from the right maxillary radiolucency revealed a radicular cyst with a dense infiltrate of blastematosic cells in the capsule (Fig. 4). Immunohistochemical stainings with myeloperoxidase, CD43, CD20 and CD3 were performed. As expected, myeloperoxidase and CD43 stained tumour cells, while reactive lymphoid and plasma cells remained negative. CD20 and CD3 stained few reactive lymphoid cells but tumour cells remained negative (Fig. 5). These findings confirmed that the dense blastematosic cell population was of myeloid origin.

Chromosomal investigation revealed translocation 6;11 which is associated with poor prognosis. The patient was treated with chemotherapy according to routine procedures. However, even though the patient had a positive reaction to the therapy initially, there was a relapse after 11 months and he died two months later in spite of active therapy.

Discussion

Granulocytic sarcoma is a proliferation of immature myeloid cells in the soft tissue or bone producing a clinically evident tumour. Occurrence of granulocytic sarcoma in the oral tissues is rare, only few cases previously described.\textsuperscript{4–9} Diagnostic confirmation usually requires immunohistochemical

Figure 1  In the peripheral blood sample half of the leucocytes were blasts (top left). The bone marrow sample was diagnosed to be hyperplastic and 90% of nuclear cells were blast cells. Courtesy of Dr. Pirkko Lammi.

Figure 2  In the oral examination the maxillary and mandibular gingiva were hyperplastic and covered decayed teeth.

Figure 3  In the panoramic tomography a diffusely bordered radiolucency in the apical area of residual roots of right maxillary canine and lateral incisor were found. Radioluencies were further found in the marginal border of left maxilla and in the mandible, all related to residual roots.
stainings. Often patients with myeloid sarcoma without concomitant symptoms of leukaemia are initially misdiagnosed.³

In this case a 59-year old male was diagnosed as having acute myeloid leukaemia. He had a poor dental status with residual roots combined with radiolucencies simulating inflammatory processes. The clinical and radiographic presentation of these lesions was radicular cysts. Histological examination revealed blastematotic cells in the connective tissue capsule in one of the cysts. The diagnosis of granulocytic sarcoma was confirmed using immunohistochemical stainings, which confirmed the tumour cells as myeloid.

A granulocytic sarcoma may occur simultaneously with a typical blood and bone marrow pattern of acute myeloid leukaemia or may antedate leukaemia by many months or years.¹ It may also be the first evidence of a relapse in a patient with acute myeloid leukaemia.¹ In this case, two days prior to the operation of the maxillary region, in the peripheral blood sample half of the leucocytes were blasts. One day prior to the maxillary operation, the bone marrow sample was diagnosed to be hyperplastic and 90% of nuclear cells of the bone marrow were blast cells. Diagnoses of the systemic disease and myeloid sarcoma in the maxilla were made almost at the same time.

In the differential diagnosis the most important diseases to be considered are non-Hodgkin lymphomas of the lymphoblastic type, Burkitt lymphoma, large-cell lymphoma

Figure 4  A biopsy revealed radicular cyst with a dense infiltrate of blastematotic cells in the capsule.

Figure 5  Myeloperoxidase (up left) and CD43 (up right) stained tumor cells while reactive lymphatic and plasma cells remained negative. CD20 (down left) and CD3 (down right) stained few reactive lymphoid cells but tumor cells remained negative.
and small round cell tumours. Immunophenotyping by immunohistochemistry for expression of myeloid associated enzymes is essential for identifying myeloid sarcoma. The importance of prompt and correct diagnosis of myeloid sarcoma has to be stressed. A local treatment for myeloid sarcoma such as radiation therapy or surgical resection has been found less effective than chemotherapy at improving the disease-free interval or disease-free survival. The occurrence of myeloid sarcoma in a patient with acute myeloid leukaemia does not alter the prognosis. Conversely, for patients with known myeloproliferative disorders, the development of myeloid sarcoma is a strongly negative prognostic factor for acute myeloid leukaemia or blast crisis. In this case the prognosis was poor based on chromosomal translocation 6;11 and the patient died after one year of the diagnosis of acute myeloid leukaemia.

In the oral region including soft tissues and mandibular and maxillary bone, inflammatory processes are common. Myeloid sarcoma is easily misdiagnosed as inflammatory infiltrate. It is essential to closely look at the nature of the cells to differentiate tumour and inflammation. Diagnostic confirmation usually requires immunohistochemical stainings. A close co-operation with the oral and maxillofacial surgeon and the pathologist is essential for the correct and prompt diagnosis.

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References