**INTRODUCTION**

Granulocytic sarcoma (GS) is a rare extra medullary solid tumour composed of immature myeloid cells. These tumours often display a greenish colour due to the enzymatic action of myeloperoxidase in the tumour cells. Hence, the term 'chloroma' was given to this lesion in 1853. GS commonly involves bone, periosteum, soft tissue, lymph node, and skin. Rare occurrences in muscle, meninges, breast, mediastinum, joints and ovary have been reported. Below-knee joint involvement in GS is unusual. We report a case of generalized cutaneous granulocytic sarcoma with ankle joint involvement which subsequently developed AML-M4.

**CASE REPORT**

A 43-year-old Pakistani man presented with a 4 months history of multiple skin nodules on the abdomen, back and legs that had gradually increased in size in September 2002. Laboratory evaluations showed normal blood counts. Peripheral blood smear, bone marrow biopsy and aspirate were normocellular. Skin biopsy showed a neoplasm composed of sheets of immature cells in the dermis and subcutaneous tissue, separating collagen bundles, and surrounding nerves and skin appendages. The tumour cells were round to avoid with vesicular nuclei and prominent nucleoli. Cytoplasm was abundant and granular. Numerous mitotic figures were present. The overlying epidermis was uninvolved by the tumour (Figure 1). Immunohistochemical staining showed that the atypical cells were positive for MPO, CD45, CD68, CD43 and negative for CD3, CD20, CD34 and CD30. This was diagnosed as a case of granulocytic sarcoma.

Initial 12 months chemotherapy (hydroxyurea) resulted in a partial response. Treatment was stopped and the patient was observed. Progression was seen in August 2005 with appearance of new skin lesions, swelling and painful ankle joint. Excision biopsy of skin nodules revealed granulocytic sarcoma with similar morphology as described previously. X-ray of the ankle joint showed lytic lesion which was consistent with bony involvement by the extramedullary myeloid neoplasm (Figure 2). Patient was treated by local radiation to the ankle joint. Peripheral blood smear showed presence of blasts and bone marrow biopsy revealed AML-M4. On induction showed a neoplasm composed of sheets of immature cells in the dermis and subcutaneous tissue, separating collagen bundles, and surrounding nerves and skin appendages. The tumour cells were round to avoid with vesicular nuclei and prominent nucleoli. Cytoplasm was abundant and granular. Numerous mitotic figures were present. The overlying epidermis was uninvolved by the tumour (Figure 1). Immunohistochemical staining showed that the atypical cells were positive for MPO, CD45, CD68, CD43 and negative for CD3, CD20, CD34 and CD30. This was diagnosed as a case of granulocytic sarcoma.

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Granulocytic sarcoma was named chloroma, because of the greenish appearance of the freshly cut surface of the tumour. This green color is due to the myeloperoxidase present in leukemic cells. The term granulocytic sarcoma is preferred as the enzyme is not present in all tumours of this type.

Granulocytic sarcoma is more common in children than adults. Although any site in the body can be involved, sites commonly involved, including: skull, paranasal sinuses, sternum, ribs, vertebra, pelvis, lymph nodes and skin. Skin is the most common presentation in association with MDS. Skin lesion in granulocytic sarcoma present as rapidly growing papulonodular, erythematous or ulcerated lesions which can be solitary, multiple or diffused and are usually found on scalp, trunk and face.

Granulocytic sarcoma may present simultaneously with AML in patients with chronic myeloproliferative diseases such as; myelofibrosis with myeloid metaplasia, hypereosinophilic syndrome, polymyelosclerosis, acute lymphoblastic leukaemia, or myelodysplastic syndrome, as the first sign of relapse of AML on maintenance chemotherapy. Local radiation may be necessary in selected cases, such as those involving the spine and as in this case, joint involvement.

Tumours of monocyes represent a similar process with a similar predilection to leukemia, mainly acute monoblastic leukaemia. Monocytic lesions react with Ab to lysozyme and CD68. Rarely myeloid lesions may be composed of erythroid and megakaryocyte precursors which may be detected by glycophorin A, hemoglobin A and factor VIII, CD41, CD61, CD31.

Blastic granulocytic sarcoma may be misdiagnosed, as reported in a previous study in up to 75% cases as non-Hodgkin's lymphoma or poorly differentiated tumour, because of lack of diagnostic features. In such cases, a broad panel of immunohistochemistry and cytochemical studies should be used.

This case demonstrates an unusual presentation of granulocytic sarcoma with extramedullary generalized cutaneous and joint involvement. The patient developed AML-M4, 3 years after initial presentation.

Granulocytic sarcoma is treated as AML with systemic chemotherapy. Local radiation may be necessary in selected cases, such as those involving the spine and as in this case, joint involvement.

**REFERENCES**