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Case Report

Rare case of a granulocytic sarcoma: cytological evaluation

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ABSTRACT

Granulocytic sarcomas (chloromas) are rare extramedullary tumors consisting of primitive granulocytic cells. They arise *de novo*, or are associated with other hematologic disorders such as acute myeloid leukemia, myelodysplastic syndrome, or myeloproliferative disorders. We report here a case of a 33-year-old man who presented with extensive skin lesions all over body, including face region. The patient is a known case of chronic myeloid Leukemia (CML). Fine needle aspiration cytology (FNAC) of skin lesion was suggestive of granulocytic sarcoma. Histological evaluation of skin biopsy revealed infiltration by atypical myeloid cells. Peripheral blood smear (PBS) examination was within normal limit. Immunohistochemistry (IHC) confirmed the diagnosis of Granulocytic sarcoma. Here we present a case of granulocytic sarcoma which was suspected on FNAC findings and which was confirmed on histochemistry and immunohistochemistry. These cases illustrate the utility of FNAC in diagnosis of granulocytic sarcoma.

Keywords: Chronic myeloid leukemia, Fine needle aspiration cytology, Granulocytic sarcoma

INTRODUCTION

Granulocytic sarcomas are rare, destructive, extramedullary tumor masses that consist of immature granulocytic cells. The term "chloroma" is derived from the Greek word *chloros* (for green). It describes that the tumor is frequently greenish color, which is due to the presence and oxidation of the enzyme myeloperoxidase in the tumor. These tumors can arise de novo or can be associated with other myeloid disorders – acute myeloid leukemia (AML) or CML, myeloproliferative, or myelodysplastic conditions.[1]

Presentation can occur prior to, in association with the underlying myeloid disorder, or upon relapse. The location of the tumor varies e.g., subperiosteal bone-skull, pelvis, ribs, sternum; lymph node; skin; gums. In this case, the granulocytic sarcoma presented in the form of varying sized skin nodules distributed all over face, neck, trunk and extremities. Such presentation is rare.[2] So far very few cases have been diagnosed on FNAC.[3-6]

CASE REPORT

A 33 years old man came with the complaints of extensive skin lesion distributed all over face, neck, trunk and extremities since two months. On examination these skin lesions comprised of multiple nodules of varying sizes. Some nodules appeared hemorrhagic and skin nodules on face led to disfigurement of face and obstruction of vision as shown in Figure 1. Some lesions have property of bleed on touch.

Past history revealed that patient was known case of CML was "philadelphia chromosome" positivity and was on imatinib therapy since two years but patient has stopped taking imatinib since last 2 months. Systemic clinical examination revealed no abnormality. X- Ray

chest, USG abdomen/CT revealed no abnormality. PBS as well as CBC findings were normal. FNAC was adviced. FNAC smears showed cell population comprised of immature cells of granulocytic series as shown in Figure 2.

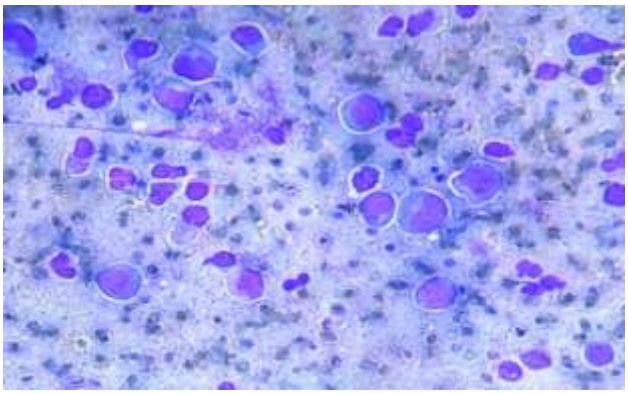

Figure 1: FNAC from face nodule show mostly metamylocytes, few blast and neutrophils.

Biopsy from skin nodule was done. Biopsy showed dermal infiltration by mononuclear cells in upper as well as deeper dermis. Skin adnexal tissue also showed mononuclear cells infiltration. IHC on skin biopsy showed that cells infiltrating dermis were found positive for cKit and MPO while CD34 was found negative. Background lymphoid cells were found highlighted by CD3 These findings confirmed the diagnosis of granulocytic sarcoma affecting skin.

Figure 2: Patients presents with multiple skin nodules over face.

DISCUSSION

The WHO has classified granulocytic sarcomas into 3 main types, depending upon the degree of maturation: 1) Blastic – composed mainly of myeloblasts 2) Immature-myeloblasts and promyelocytes 3) Differentiated-promyelocytes and more mature myeloid cells. Rarer types can consist of a monoblastic sarcoma, associated with monoblastic leukemia.[7]

In this case the patient being known case of CML though in remission, and cytology smear showing immature granulocytic series cells, suggested the diagnosis of granulocytic sarcoma. Full investigations into the morphology, immunohistochemistry, immunophenotyping, and cytogenetics are essential as the diagnosis may be inconclusive on the basis of morphology alone. The morphology of the cells can be variable.

If well differentiated, a diffuse infiltrate of granulocytic cells can be seen (often containing all stages of myeloid cell maturation). If poorly differentiated, the majority of the cells are a large and monotonous population. The nucleoli may or may not be prominent. Cytoplasm is often scanty. A high mitotic index can be seen. Tissue both from the tumor and the marrow needs to be examined as the cellularity and phenotype can differ between them.

CML can be associated with granulocytic sarcomas in a variety of ways. Most frequently, the tumor presents later in the natural history of the CML as the disease progresses. Patients in relapse after chemotherapy or stem cell transplant can also present with a granulocytic sarcoma.[8] Of note, a granulocytic sarcoma that develops with chronic phase CML confers a worse prognosis – ie, a higher risk of rapid blastic marrow transformation.[9]

Any underlying CML associated with the granulocytic sarcoma will require appropriate treatment, such as with imatinib mesylate. This targeted therapy inhibits the abnormal Bcr-Abl tyrosine kinase created by the Philadelphia chromosome abnormality. Imatinib inhibits cell growth and induces apoptosis in the affected cells, and should be used in combination with the standard AML therapy.[10]

CONCLUSION

FNAC smear can be useful in the diagnosis of granulocytic sarcoma. Cytochemistry / immunocyto-chemistry can enable us to confirm the diagnosis of granulocytic sarcoma.

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REFERENCES

1. Jenkins CI, Sorour Y. Case Report: A Large Extramedullary Granulocytic Sarcoma as the Initial Presenting Feature of Chronic Myeloid Leukemia. MedGenMed. 2005;7:23.
2. Campidelli C, Agostinelli C, Stitson R, Pileri SA. Myeloid Sarcoma- Extramedullary Manifestation of

Myeloid Disorders. Am J Clin Pathol. 2009;132:426-7.
3. Kumar PV: Soft tissue chloroma diagnosed by fine needle aspiration cytology. A case report. Acta Cytol. 1994;38:83-6.
4. Pettinato G, De Chiara A, Insabato L, De Ren-zo A. Fine needle aspiration biopsy of a granulocytic sarcoma (chloroma) of the breast. Acta Cytol. 1998;32:67-71.
5. Dunphy CH, Katz RL, Fanning CV, Dalton WT. Leukemic lymphadenopathy: Diagnosis by fine needle aspiration. Hematol Pathol. 1989;3:35-44.
6. Raab SS, Silverman JF, McLeod DL, Geisinger KR. Fine-needle aspiration cytology of extra-medullary hematopoiesis (myeloid metapla-sia). Diagn Cytopathol. 1993;9:522-9.
7. Jaffe ES, Harris NL, Stein H, Vardimann JW, editors. World Health Organisation Classification of Tumours-Tumours of Haematopoietic and Lymphoid Tissues. 2001:105.
8. Martel L, Reddy K, Greco M, Tuscano M, Richman M, Wun T. Isolated cavernous sinus extramedullary relapse of chronic myelogenous leukemia following allogeneic stem cell transplant. Ann Hematol. 2002;81:108-10.
9. Mahendra P, Ager S, Bedlow AJ, Bloxham DM, Green AR, Marcus RE. Two unusual neurological presentations of granulocytic sarcoma in Philadelphia positive chronic myeloid leukaemia. Leuk Lymphoma. 1994;15:351-5.
10. Druker BJ1, Talpaz M, Resta DJ, Peng B, Buchdunger E, Ford JM, et al. Efficacy and safety of a specific inhibitor of the BCR-ABL tyrosine kinase in chronic myeloid leukemia. N Engl J Med. 2001;344:1031-7.

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