GRANULOCYTIC SARCOMA PRESENTING AS A BONE TUMOR

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ABSTRACT

Granulocytic sarcomas (chloromas) are very rare tumors that occur outside hematopoietic areas, it consists of primitive granulocytic cells. They arise de novo, or may be associated with other hematologic disorders such as acute myeloid leukemia, myelodysplastic syndrome, or myeloproliferative disorders. We report here a case of a 32-year-old man who presented with drowsiness, paraplegia, fever and bone pains for the last 3 months. There was equivocal evidence for caries spine in clinical history, examination and upon routine investigations. X-rays and MRI spine showed anterior wedge fractures of D_{10} & L_1. Technetium bone scan pointed to hot areas in the pelvis and spine. The biopsy taken from right Iliac crest showed it to be a granulocytic sarcoma. Bone marrow examination showed a normal picture.

Key words: Granulocytic sarcomas, Myeloblasts, Extramedullary.

INTRODUCTION

Granulocytic sarcomas (chloromas) are rare tumors occurring outside hematopoietic areas consisting of primitive granulocytic cells.[1] They arise de novo, or are associated with other hematologic disorders such as acute myeloid leukemia, myelodysplastic syndrome, or myeloproliferative disorders.[2]

CASE REPORT

A 32 year old male presented to the out patient department with inability to walk, off & on fever, bone pains for the last three months and altered sensorium for the last few days. He had been suffering from weakness of legs during this period but in the last few days the fever became high grade with ensuing loss of consciousness which made his attendants bring him to the hospital. He had got treatment from many doctors till his admission. History revealed a gradual weight loss in last six months. On examination the patient was drowsy, pale and emaciated. His lower limb showed absent muscle tone, the reflexes could not be elicited, plantar reflexes were non specific and there were no signs of meningeal irritation. There were no other findings on a thorough physical examination.

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The blood count showed a normal total leukocyte count accompanied by neutrophil predominant (88%) picture with a high ESR. Biochemical studies showed a normal liver and kidney function along with a euglycemic status ruling out metabolic reasons of drowsiness. Peripheral blood picture was unremarkable (Table I). Keeping in view the steady weight loss over the last six months, paraplegia and bone pains which were especially severe in the spine. X-rays were taken of the dorsolumbar region which revealed anterior wedging of the D_{10} & L_1 vertebrae. The MRI showed an infiltrative process eroding the said vertebral bodies compressing the spinal cord, comparable with the clinical finding of paraplegia. A $Tc_{99}m$ bone scan at this stage showed hot areas in the cervical and left iliac regions. Radiological surveillance of the area showed erosion of the C_2 body. (Table II & Figure I)

Table: I
Laboratory findings

Complete blood picture	
Hb	10.4g/dl
WBC	11800
Neutrophils	88%
Basophils	2%
Lymphocytes	07%
Monocytes	03%
ESR	85 in 1st hour
Platelets	720000 per cmm
Clotting Profile	
PT	14/13
APTT	35/33
Biochemical Analysis	
S. Calcium	10.5 mg/dl
S. Phosphorus	3.9 mg/dl
PTH	< 3 pg/ml (Normal)
Bence Jones Proteins	Negative

Table II: Radiological and histopathological findings

MRI Spine	Multiple vertebral body infiltrating lesions, one in the lower dorsal spine is causing compression on cords and nerve roots.
Bone marrow biopsy	Normal
Bone imaging:	Pathological bone disease in D_{10} & L_1 vertebrae. Could be metastatic
FNAC: (Right iliac bone lesion)	Granulocytic sarcoma/chloroma
Histopathology:	Inconclusive
MRI brain:	Demyelinating plaques in white matter of both cerebral hemispheres. Bone eroding mass (metastatic) involving the body of c-2 vertebra

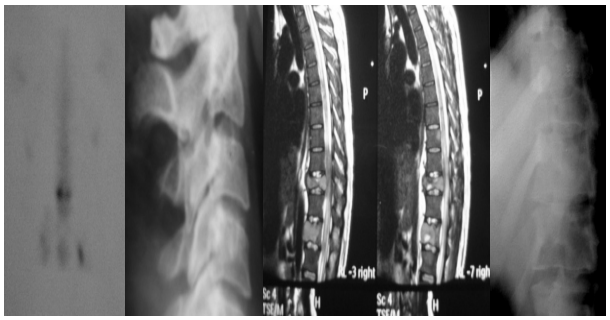

Figure: I
Radiological Pictures

A fine needle aspiration cytology was done from the right iliac crest which showed granulocytic sarcoma picture. However the histopathological study that followed was inconclusive. The MRI brain showed demyelinating plaques in the white matter of both the cerebri and an erosive mass in the body of C_2.(Table II)

DISCUSSION

Granulocytic sarcomas are rare, destructive, extramedullary tumor masses that consist of immature granulocytic cells.[1] The term "chloroma" is derived from the Greek word chloros (for green) because the tumor is frequently Greenish in color, which is due to the presence and oxidation of the enzyme myeloperoxidase in the tumor.[2,3] They occur in 2.5-9.1% of patients with acute myelogenous leukemia and five times less frequently in patients with chronic myelogenous leukemia (CML).[1,2] Granulocytic sarcomas have approximately the same rate of occurrence in both sexes. Children are more often affected than adults; 60% of patients are younger than 15 years Old. These tumors can arise de novo or can be associated with other myeloid disorders – acute myeloid leukemia (AML) or CML, myeloproliferative, or myelodysplastic conditions.[2,3] Presentation can occur prior to, in association with the underlying myeloid disorder, or upon relapse.[4] In some cases the tumor occurs in isolation without any other associated hematological malignancy.[1] The location of the tumor varies – e.g., subperiosteal bone skull, pelvis, ribs, sternum; lymph node; skin; gums. In this case, the granulocytic sarcoma was multi focal giving rise to spinal symptoms due to local destruction. The paraplegia resulted from the collapse of the vertebral bodies which mimicked caries spine in the presence of a high ESR (85mm). The involvement of the cervical spine was asymptomatic. We were not able to find, any cases that were similar in characteristics (age and diagnosis) to the patient in the case presented here. The right iliac bone involvement and the lack of local osteopenia the hallmark of tuberculous bone disease lead to the suspicion of a metastatic process. The non invasive bone scan and MRI lead to a clearer picture.

WHO has classified granulocytic sarcomas into 3 main types, depending upon the degree of maturation.[2]

Blastic
– composed mainly of myeloblasts
Immature
– myeloblasts and promyelocytes
Differentiated
– promyelocytes and more mature myeloid cells

Rarer types can consist of a monoblastic sarcoma, associated with monoblastic leukemia. Clinically and histologically, the diagnosis needs to be distinguished from the main differentials – ie, Hodgkin lymphoma, Burkitt's lymphoma, large cell lymphomas, and small round blue cell tumors, such as neuroectodermal tumors.[5] These sarcomata are

very sensitive to focal irradiation or chemotherapy; they generally resolve completely in less than 3 months, although they recur in approximately 23% of patients. Patients with granulocytic sarcomata are frequently asymptomatic: 50% of cases are diagnosed only at autopsy. These tumors can involve any part of the body, either concurrently or sequentially. They often occur in multiples and preferentially involve orbits and subcutaneous tissue, but they may also occur in paranasal sinuses, lymph nodes, bone, the spine, the brain, pleural and peritoneal cavities, the breasts, the thyroid, salivary glands, the small bowel, the lungs, or various pelvic organs.[1,2]

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5. Adult Primitive Neuroectodermal Tumor (PNET) of the

Hazrat Ali (ÑÖí ÇááÀ ÊÚÇáí ÚäÀ) Said:

Whoever wants to be a leader should educate himself before educating others. Before preaching to others he should first practice himself. Who ever educates himself and improves his own morals is superior to the man tries to teach and train others.