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Granulocyte sarcoma presenting as acute Paraplegia and Proptosis

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Abstract :

Paraplegia due to a spinal cord epidural mass is an extremely rare presentation of undiagnosed leukemia. Paraplegia with co-existing proptosis as a initial presentation due to chloroma is extremely rare entity. Here, we report a case of 16 year old boy who presented to Emergency Department with acute paraplegia and urine retention due to multiple cervical, thoracic and sacral epidural masses and progressive bilateral proptosis due to extra-conal masses, as an initial presentating manifestation of acute myeloid leukemia. Granulocytic sarcoma or chloroma should be considered in the differential diagnosis of an epidural mass. These are rare extramedullary tumor-like proliferation of myelogenous precursor cells that may de novo precede acute leukemia or coincide with the first manifestation or relapse of acute myeloid leukemia.

Key-words: *Granulocytic sarcoma, compressive myelopathy, acute myeloid leukaemia*

Introduction

Granulocytic sarcoma (GS) or chloroma has been defined as an extramedullary tumour mass consisting of myeloid blasts with or without maturation.[1,2]Paraplegia due to chloroma is an extremely rare presenting neurological manifestation in myeloid leukemia.[1] Chloromas are most frequently seen in acute myeloid leukemia (AML).[1-4] Also, it may signal the onset of accelerated phase of chronic myeloid leukemia or blastic transformation of a myeloproliferative disorder. GS should be considered in the differential diagnosis of an epidural mass in patients with or without leukaemia as Granulocytic Sarcoma represent a diagnostic challenge, particularly those occurring in patients without evidence of systemic disease.[1]

Case Report

A 16 year old boy presented wit low backache following a fall from a height of 4 feet, two weeks ago. He was apparently normal for 2-3 days following fall, after which he developed progressive left eye swelling (Figure 1), weakness of lower limbs, retention of urine and fever since 2 days. On examination, patient had pallor and was thin built left eye showed proptosis, fundus was normal. There was spinal tenderness at lumbosacral region and mild diffuse tenderness of abdomen. Lower limbs had grade 3 power, with absence of tendon reflexes and bilateral extensor plantar response. Cranial nerves, sensory system were unremarkable, other systems were normal

Hematological investigations revealed Hb : 7.2 g/dl, RBC 2.5 million/mm3,TC : 11200 cells/mm^3, with a differential count of N 41%, L-28% , E-01%, M-30%, ESR 120 mm/hr, Platelet count: 51,000/mm^3, PT/INR - Mild hepatosplenomegaly with minimal ascites noted on the ultra sound. Ascitic fluid of Gram stain, AFB stain, cultures, and cytology for malignant cells were negative. Peripheral smear showed dimorphic anemia, thrombocytopenia and myeloblasts suggestive of subleukemic leukemia(figure 2)

History of trauma causing proptosis and paraparesis with radiograph failed to demonstrate any compression fracture in dorso-lumbar spine. chest x-ray was normal. chest x-ray was normal. CT Brain plain showed intra orbital mainly extra-conal masses seen bilaterally. (Figure 3). MRI Orbit T1 weighted pre contrast images (right) done in the sagittal plane revealed extra-conal mass which is isointense to brain, on post contrast imaging (left) there is heterogeneous enhancement of the extra-conal mass.(Figure 4).MRI Spine showed Multiple midline fusiform extra-dural masses which are iso-intense to cord on T1W images seen extending from C3-C7, D3-D5, D11-D12 & S1-S2 are seen posteriorly in the

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spinal cord causing significant compression of the spinal cord.(Figure 5). Considering the possibility of spine and orbital granulocytic sarcoma associated with leukaemia, Bone marrow aspirate was done which confirmed the diagnosis of acute myeloblastic leukaemia (AML) of the French-American-British M2 subtype.(Figure 6) and he was was started on local radiotherapy and chemotherapy

Discussion

Granulocytic Sarcoma (also termed myeloid sarcoma or chloroma) is a rare malignant neoplasm resulting from the extramedullary proliferation of myeloid blasts with or without maturation.[12] Macroscopically, chloromas are green in appearance due to the myeloperoxidase in the leukaemia cells and fades when exposed to air.[1-8] Granulocytic sarcoma occurs in 3% to 9% of AML.[7] Rarely chloroma can occur alone without peripheral blood or bone marrow evidence of leukaemia.[1] Moreover, 66% -88% of patients with an isolated granulocytic sarcoma will develop AML at a mean of 9-11 months after diagnosis.[4] Granulocytic sarcomas are generally seen in the ribs, sternum, pelvis and orbital bones as well as in the soft tissues, lymph nodes, skin, and gums.[6-12] Involvement of the central nervous system (CNS) is rare, and spinal cord compression by granulocytic sarcoma is even more rare.[2-9, 6-12.]

In our patient, paraplegia and proptosis due to chloroma as the initial presenting feature of AML is very rare and not mentioned in any literature. In the present case, the GS occurs with peripheral blood evidence of leukaemia. The correct diagnosis of GS is obtained in only about 50% of non-leukaemic patients due to its rareness and the histological and radiological similarities to malignant lymphoma.[12-14] Chloroma should be considered in the differential diagnosis of any epidural mass in patients with or without leukaemia.[1] MRI usually shows epidural mass lesion compressing the cord, isointense with cord in T1, and hyperintense in T2-weighted images. The differential diagnosis includes epidural abscess, metastasis, and lymphoma deposits. Lymphoma usually shows epidural extension of tumor directly from vertebral body and inhomogeneous hyperintense signals in T2 images of vertebral body due to marrow infiltration, which is not seen in chloroma. CSF cytology for malignant cells will be negative in extradural compressive myelopathy due to chloroma as in our case.

Optimal therapy for patients with GS is not yet well defined. GSs are usually radiosensitive and are often treated with local radiotherapy and chemotherapy.[3,5] Newly diagnosed patients with isolated GS usually treated with aggressive chemotherapy as if they have acute myelogenous leukaemia, cures are not attained with radiation therapy alone[5] Surgery is generally preferred for cases of acute spinal cord compression in cases without systemic evidence of leukaemia.[3] Early diagnosis followed by appropriate combined chemotherapy and radiation may obviate surgical intervention.[1]

Figure 1 : Left eye Proptosis

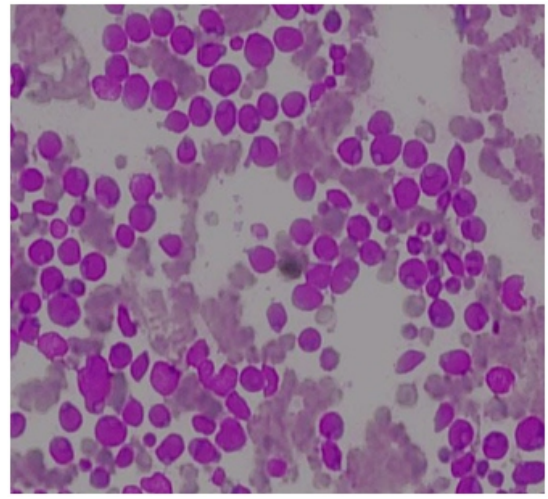

Figure 2 : Peripheral smear predominantly blast cells with multiple nucleoli

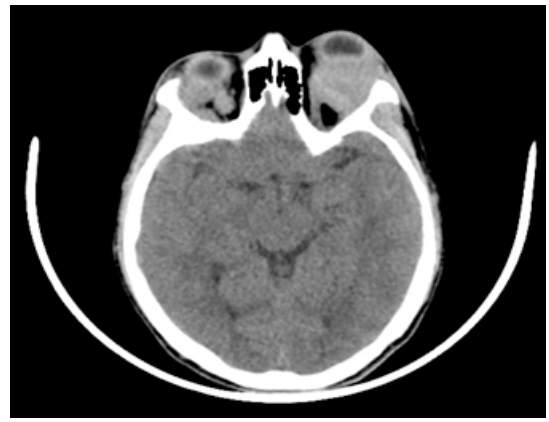

Figure 3 : Axial CT images showing bilateral extra conal masses significantly large on the left side causing proptosis

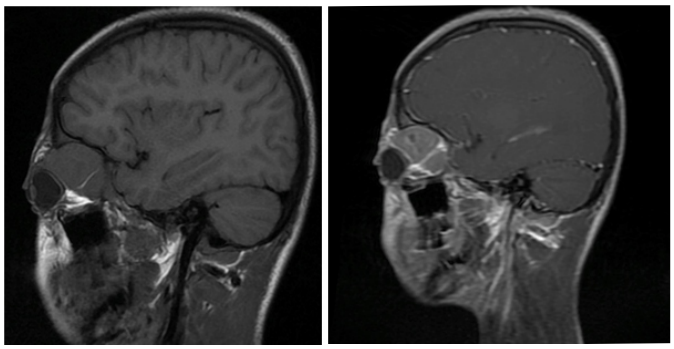

Figure 4 : T1 weighted pre contrast images (right) done in the sagittal plane revealed extra-conal mass which is isointense to brain. On post contrast imaging (left) there is heterogeneous enhancement of the extra-conal mass.

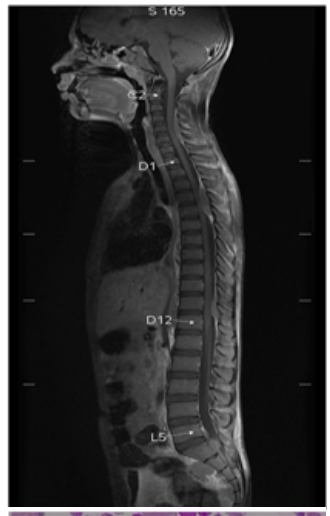

Figure 5 : MRI SPINE shows Multiple midline fusiform extra-dural masses which are iso-intense to cord on T1W images seen extending from C3-C7, D3-D5, D11-D12 & S1-S2 are seen posteriorly in the spinal cord causing significant compression of the spinal cord.

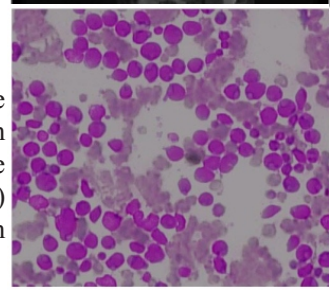

figure 6 : Bone marrow aspirate showed blast cells cells which confirmed the diagnosis of acute myeloblastic leukaemia(AML) of the French-American-British M2 subtype.

References

1. Amalraj P, Syamlal S: Unusual case of paraplegia. Ann Indian Acad Neurol. 2009 ; 12(3): 188- 90.
2. Balleari E, Panarello S, Capello E, et al: Granulocytic sarcoma: an unusual cause of spinal cord compression. Int J Clin Oncol 2007;12: 234-7.
3. Buckland ME, Scolyer RA, Donellan MB, Harper CG: Spinal chloroma presenting with triplegia in aleukaemic patient. Pathology. 2001;33: 386-9.
4. Chevallier P, Mohty MB, Lioure B, et al: Allogeneic haematopoietic stem-cell transplantation for myeloid sarcoma: a retrospective study from the SFGM-TC. J Clin Oncol 2008, 26: 1-4.
5. Choudhari R, Bhattacharya M, Mishra P, Mahapatra M, Kumar R: Granulocytic sarcoma of spine: An unusual initial presentation of acute myeloid leukaemia. J Asoc Physicians India. 2006; 4: 897-9.
6. Cui Y, Zhou JL, Wu JH, Zhang JZ: Synchronous granulocytic sarcoma of the breast and spine: a case report and review of literature. Chin Med J (Engl). 2008 Sep 20; 121(18): 1854-6.
7. Ghalaut PS, Jindal S, Jeevan O: Granulocytic sarcoma as initial presentation of acute myeloid leukaemia. J Asoc Physicians India. 2005; 53: 828-30.
8. Landis DM, Aboulafia DM. Granulocytic sarcoma: An unusual complication of aleukaemic myeloid leukaemia causing spinal cord compression. Leuk Lymphoma. 2003; 44: 1753-60.
9. Neiman RS, Barcos M, Berard C, et al: Granulocytic sarcoma: a clinicopathologic study of 61 biopsy cases. Cancer 1981, 48: 1426-37.
10.Novick SL, Nicol TL, Fishman EK: Granulocytic sarcoma (chloroma) of the sacrum: initial manifestation of leukaemia. Skeletal Radiol. 1998; 27: 112-4.
11. Park HJ, Jeong DH, Song HG, Lee GK, Han GS, Cha SH, et al: Myeloid sarcoma of both kidneys, the brain, and multiple bones in a nonleukaemic child. Yonsei Med J 2003; 44: 740-3.
12. Serefhanoglu S, Goker H, Aksu S, Buyukasik Y, Sayinalp N, Haznedaroglu IC and Ozcebe Ilhami O: Spinal myeloid sarcoma in two nonleukaemic patients. Intern Med 2010, 49: 2497-8
13. Traweek ST, Arber DA, Rappaport H, Brynes RK: Extramedullary myeloid cell tumours: An immunohistochemical and morphologic study of 28 cases. Am J Surg Pathol 1993, 17: 1011-9.
14. Williams MP, Olliff JF, Rowley MR: CT and MR findings in parameningeal leukaemic masses. J Comput Assist Tomogr 1990;14: 736- 42.
15. Seo HY, Kim SJ, Choi JW, Lee JB, Kang CH. Unusual relapse of acute myeloid leukemia; Granulocytic sarcoma presenting as acute paraplegia. Ann Hematol. 2006;85:196–7.
16. Mostafavi H, Lennarson PJ, Traynelis VC. Granulocytic sarcoma of the spine. Neurosurgery. 2000;46:78–83.

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