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MYELOID SARCOMA OF PARAVERTEBRAL REGION- A CASE REPORT

Pathology

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ABSTRACT

Myeloid sarcoma is a tumor of myeloid blasts, mature or immature, in a site other than bone marrow, causing tissue architectural effacement and often occurs along with myeloid neoplasm of marrow with variable temporal relationships. Primary myeloid sarcoma of paravertebral region is a rare tumor. A 60-year-old female presented with acute onset neurological deficit with bladder and bowel dysfunction and bilateral lower limb weakness. Initially thought of as a neural tumor of paravertebral region, histopathological examination, followed by immunohistochemistry proved it to be myeloid sarcoma.

KEYWORDS

Myeloid sarcoma, Paravertebral, Immunohistochemistry

INTRODUCTION

Myeloid sarcoma is a tumor mass of myeloid origin, occurring at a site other than bone marrow, with resultant alteration in tissue architecture.[1] It occurs preceding, concurrently or following myeloid neoplasms, or rarely occur as isolated lesion, thus called primary myeloid sarcoma. Histopathological examination followed by immunohistochemistry forms the cornerstone of diagnosis in such primary cases, often due to the rarity of the lesion, and unusual location it presents.

Case Presentation

A 60-year-old female manual labourer presented with acute onset of urinary retention, constipation, and weakness of bilateral lower limbs. She has been having back pain for past 1 month and weakness and decreased sensation of bilateral lower limbs for past 10 days. There was no history of trauma, or known comorbidities.

On examination, GCS was E4M5V6, PEARL; Pulse rate- 92/min;BP - 140/80 mm of Hg; O2 Saturation- 96%; Afebrile. Higher mental functions and cranial nerve examination were within normal limits. Bulk and tone were normal in all limbs. Power was 0/5 in both lower limbs and 5/5 in both upper limbs. Sensations were lost below D12 dermatome. Routine blood counts, liver function tests, renal function tests, PT-INR, APTT and viral markers were all unremarkable. Urine culture revealed mixed bacterial growth.

MRI scan of lumbar spine was carried out and showed moderately enhancing, altered signal intensity mass lesion in the left paravertebral region, extending to left neural foramina, posterior peridural space and left posterior paraspinal muscles involving erector spinae muscles, at D9-D10-D11-D12 vertebrae region. Along the epidural space, the lesion was seen causing mass effect on spinal cord, in the form of compression and displacement anteriorly. The lesion measured approximately 9cm in craniocaudal direction and 4.5cm in anterioposterior direction. There was no bony erosion. Possibility of a neurogenic tumour with left paravertebral and left posterior paraspinal muscle extension was made, with a differential diagnosis of lymphoma.

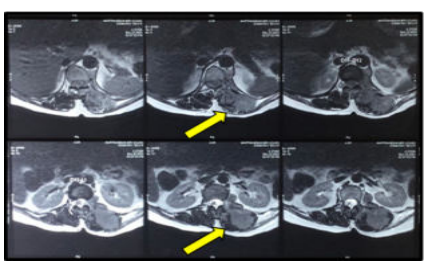
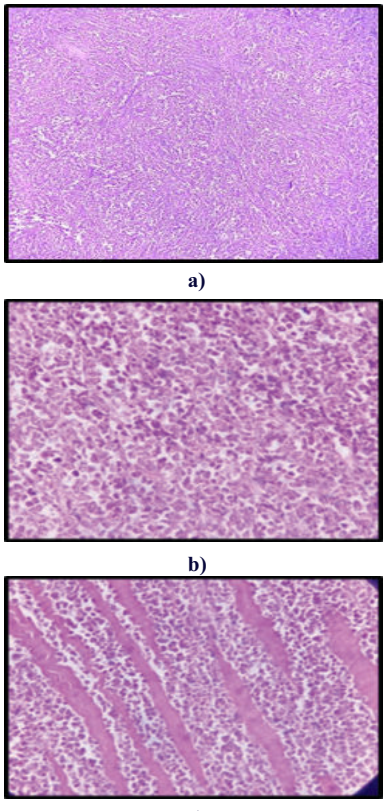


Figure 1: MRI lumbar spine

USG guided FNAC did not reveal any significant results. CBNAAT for Mycobacterium tuberculosis from the lesion was negative. Patient was posted for surgery and D9-D12 laminectomy and biopsy under general aneasthesia was done. Frozen section analysis was done and was reported as small round cells, possibly lymphoproliferative disorder.

On biopsy, we received multiple grey brown tissue bits aggregate measuring 19 x 6 x 2 cm. Microscopy showed an infiltrating neoplasm arranged in sheets, singly and along blood vessels. Individual cells were medium to large sized, with scant cytoplasm, high N/C ratio, and round vesicular nuclei with fine chromatin and prominent nucleoli. Also noted were a few cells with irregular nuclear membranes. There was brisk mitotic activity and there were scattered apoptotic bodies. The neoplasm was seen infiltrating into adjacent muscle.



a)

b)

c)

Figure 2: H & E images a) Neoplasm is arranged in a diffuse pattern. b)

Cells are medium to large sized with scant cytoplasm high N/C ratio and round vesicular nuclei. c) Neoplasm is seen infiltrating adjacent muscle.

Immunohistochemistry was carried out and showed, diffuse and strong positivity for CD34, MPO and CD117 in the neoplastic cells. LCA was focally positive. Tdt, CD3, CD20, CD138, S100, and CD68 were negative. With the above findings a possibility of myeloid sarcoma was considered. Haematological work up was absolutely indicated.

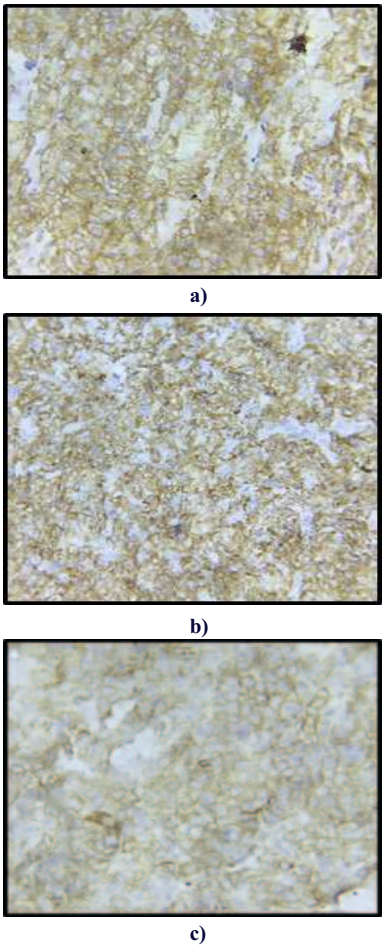


Figure 3: Neoplastic cells show diffuse strong positivity for a) CD117, b) MPO, c) CD34.

Post operative period was uneventful and the patient was referred to Regional Cancer Centre (RCC) for further management. Slide review was done at RCC, and the diagnosis remained unchanged. They had carried out few additional immunohistochemical markers, CD30 and CD5 negative, ERG -positive, CD33- positive (patchy and weak). Peripheral smear was unremarkable, with normocytic normochromic RBCs, normal WBC count and adequate platelet counts. Bone marrow aspirate was cellular and showed mild lymphocytosis and eosinophils only. Bone marrow trephine biopsy showed trilineage hematopoesis with mild eosinophilia. PCR Assay for BCR-ABL fusion transcript showed no fusion. Cytogenetic study in bone marrow revealed no analysable metaphases.

MRI spine was done at RCC which showed residual lesion. Patient underwent 10 cycles of radiotherapy. Post treatment she has begun to perceive pain in both lower limbs and appreciates touch. She is advised to undergo further chemotherapy in local hospital.

DISCUSSION

Myeloid sarcoma is a neoplasm of mature or immature myeloid blasts, occurring at an anatomical site other than bone marrow, with effacement of tissue architecture.[1] This rare neoplasm was first described by King, and he called it chloroma owing to the distinctive green appearance due to the presence of myeloperoxidase enzymes.[2] It occurs over a wide age range (1 month to 89 years) with a mean patient age of 56 years. It has slight male preponderance. Most common sites of involvement are skin, lymph nodes, gastrointestinal tract, bone, soft tissue etc.

It often occurs preceding, concurrently, or following myeloproliferative disorder.[3] In one quarter of cases it occurs in the absence of any myeloid neoplasms,[1] and is termed as primary myeloid sarcoma. The incidence of primary isolated MS is approximately 2/1,000,000 in adults, and 0.7/1,000,000 in children.[4,5]

Isolated primary MS of paravertebral region is rare. In our case, the patient had left paravertebral mass lesion which caused mass effect and compression on the spinal cord leading to acute on chronic symptoms. Initially thought of as a neurogenic lesion by radiology, the diagnosis was made through biopsy and histopathological examination. Histopathology alone doesn't prove helpful in making a diagnosis of myeloid sarcoma because non Hodgkin lymphoma, undifferentiated carcinoma and large cell lymphoma are close differential diagnoses.[6–8] On immunohistochemistry, these tumor cells lack specific B and T cell markers. Tumors with immature myeloid profile express CD33, CD34, CD68 and KIT (CD117). TdT, MPO, and CD45 show inconsistent staining. Promyelocytic cases express MPO and CD15, but lack CD 34 and TdT. Myelomonocytic, monoblastic, rare erythroid, and megakaryoblastic MS express lineage specific markers. In more than half of the cases, CD99 is positive with no association with a specific subtype.[1]

Compared to AML, non AML MS may have a better survival; however, MS is often associated with a poor prognosis.[9–11] Mean survival time for these patients range between 2.5 and 22 months. Untreated patients have even worse prognosis.[12] It is at this point that accurate diagnosis and prompt management becomes of utmost importance. Age,sex, anatomical site involved, type of presentation, therapy relatedness, histological features, immunophenotype or cytogenetic findings have no impact on clinical behaviour and response to therapy.[1] Surgery or radiotherapy may be the first line of treatment in patients where debulking is considered for symptomatic relief. Surgery may be followed by chemotherapy, as was done in our patient.

CONCLUSIONS

Isolated myeloid sarcoma in the absence of myeloid neoplasms is a rare entity. Diagnosis of isolated or primary myeloid sarcoma becomes difficult, in instances of unusual locations, absence of an associated bone marrow disease and non-specific symptoms. Prompt identification and diagnosis is essential because it can be easily mistaken for lymphoma or undifferentiated carcinoma, and because untreated cases are known to have worse prognosis when compared to cases who have received prompt treatment.

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