

Isolated extramedullary myeloid sarcoma causing cauda equina compression in a young male

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ABSTRACT

We report a case of a 22-year-old young male presenting with radicular pain of lower limbs, with urinary retention and erectile dysfunction due to cauda equina compression by a pre-sacral mass, which was shown to be an extramedullary myeloid sarcoma on histopathology and immunohistochemistry. However, the patient did not have any evidence of acute myelogenous leukaemia either in peripheral blood or bone marrow. He responded favourably to treatment by local external beam radiotherapy and systemic chemotherapy using the 3 + 7 protocol. Isolated extramedullary myeloid sarcoma causing cauda equina compression is extremely rare, with very few case reports published earlier. Our case also emphasizes the importance of aggressive combined-modality treatment in such patients to achieve durable remission and preserve reasonable quality of life.

Key words: Cauda equina compression, extramedullary myeloid sarcoma, pre-sacral mass

INTRODUCTION

Myeloid sarcoma is a solid, extramedullary tumour composed of leukaemic myeloblasts or immature myeloid cells.^[1] In the 2002 World Health Organization classification of myeloid neoplasms, the terms “granulocytic sarcoma” and “chloroma” that were commonly used to refer to such tumours were replaced with “myeloid sarcoma”.^[2] Myeloid sarcoma involves almost all tissues, including those of the skin, lymph nodes, spine, orbit, and bone.^[3] Occasionally, myeloid sarcoma precedes the development of systemic disease by weeks to years. Myeloid sarcomas may rarely affect the spinal canal, causing spinal cord or cauda equina compression, of which very few cases have been reported. We report the rare occurrence of pre-sacral myeloid sarcoma in an aleukaemic young male presenting with features of cauda equina compression.

CASE REPORT

A 22-year-old previously healthy, young male was admitted with a history of radicular pain in the posterior aspect of both lower limbs along with numbness over the same distribution for 4 months; difficulty in micturition, with urinary retention, for 3 months; and loss of spontaneous early morning erection for 2 months, without any preceding history of trauma, fever, tuberculosis, bone pain, or unexplained weight loss. Physical examination showed mild pallor and diffuse tenderness over the sacral area in the absence of lymphadenopathy or hepatosplenomegaly. On neurological examination, his higher mental function, cranial nerves, and upper limb function were normal. The muscle tone of both lower limbs was normal, with good power in all major muscle groups, except the bilateral extensor hallucis longus, which were significantly weak (3/5). There was no fasciculation. Pain and temperature sensations were diminished over the S₁-S₄ sensory dermatomes, with preserved posterior column sensations in both lower limbs. Abdominal reflex and cremasteric reflex were normal. Anal reflex and bulbocavernosus reflex were absent. Plantar reflex was bilaterally unresponsive. All deep tendon reflexes were normal except ankle jerk, which was bilaterally absent. The straight leg raising test was bilaterally normal (90°). Tests for cerebellar function were normal bilaterally.

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Initial blood reports showed normocytic normochromic anaemia, normal serum electrolytes, and normal urine microscopy. HIV serology was negative. Ultrasound examination of abdomen was normal, without any intra-abdominal or pelvic lymphadenopathy. Chest radiograph showed absence of mediastinal widening, with a normal lung parenchyma. Mantoux test was negative. Magnetic resonance imaging of the lumbosacral spine showed a long-segment epidural mass with clumping of the nerve roots of the lower cauda equina at the L₅-S₂ levels, with a large pre-sacral soft tissue mass [Figure 1]. Computed tomography-guided core cut biopsy of the sacral mass on histopathological examination showed bundles of muscle fibres infiltrated by a tumour composed of round or oval cells with hyper-chromatic nuclei and occasional eosinophilic myelocytes suggestive of a malignant small, round, blue cell tumour [Figure 2]. On immunohistochemistry the tumour cells expressed leukocyte common antigen, c-kit, CD34, TdT (focally), myeloperoxidase, and CD43, and were

immunonegative for CD20, CD3, CD30, and cytokeratin, which was consistent with a diagnosis of an extramedullary myeloid cell tumour [Figures 3 and 4]. However, bone marrow aspirate and subsequent biopsy did not show evidence of leukaemic infiltration and differentiation, and maturation of all three series was normal. So the patient was diagnosed to have an isolated pre-sacral, extramedullary myeloid cell tumour in the absence of concomitant acute myeloid leukaemia (AML).

In view of the neurological symptoms, especially radicular pain due to cauda equina compression, local external beam radiotherapy (EBRT) was started urgently. A single direct posterior portal with a Co⁶⁰ beam was used, and the dose was prescribed at a depth of 6 cm. The total dose prescribed was 24 Gy in conventional fractionation (2 Gy per fraction, single fraction per day, five fractions per week). The patient was relieved of his distressing radicular

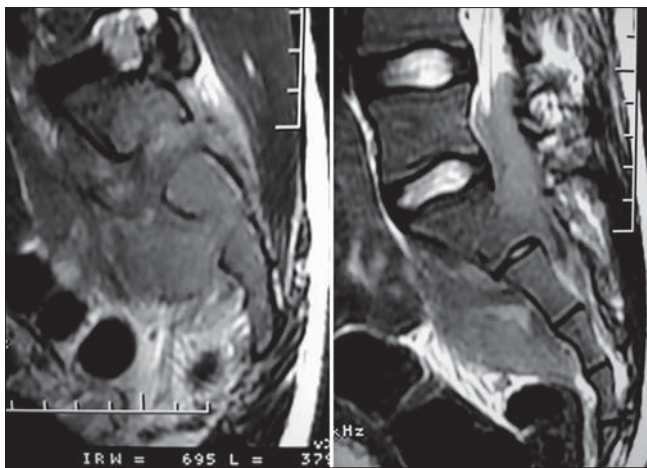


Figure 1: Magnetic resonance imaging of the lumbosacral spine showing a long-segment epidural mass with clumping of the nerve roots of the lower cauda equina at the L5-S2 levels, with a large pre-sacral soft tissue mass

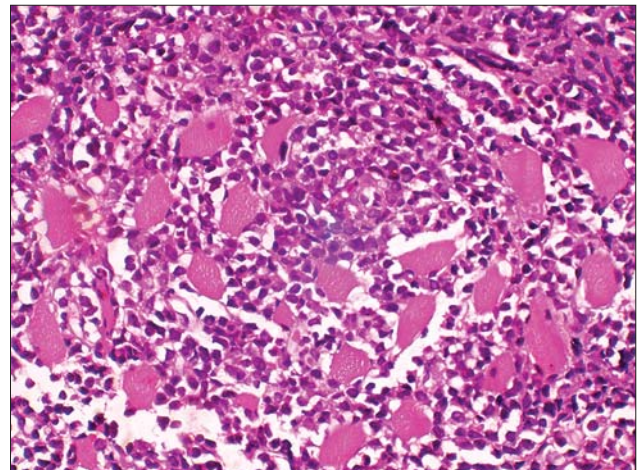


Figure 2: A photomicrograph (H and E, x400) showing bundles of muscle fibres infiltrated by a tumour composed of round or oval cells with hyper-chromatic nuclei and occasional eosinophilic myelocytes suggestive of a malignant small, round, blue cell tumour

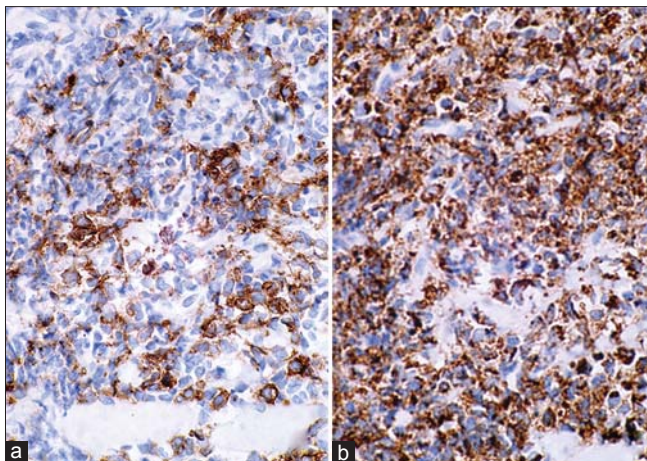


Figure 3: Immunohistochemical staining showing (a) c-kit positivity and (b) myeloperoxidase positivity

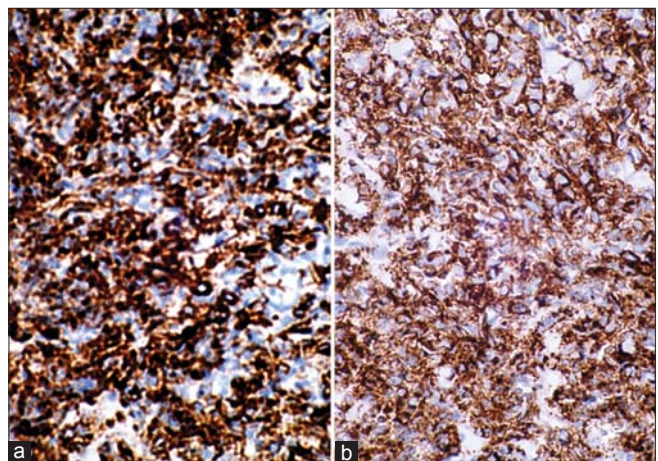


Figure 4: Immunohistochemical staining showing (a) CD34 positivity and (b) CD43 positivity

pain within a few days of completion of EBRT, but much improvement of his bladder control was not noted. Thereafter, he was treated by the standard chemotherapy protocol for remission induction in AML, which consists of 45 mg/m² Inj. daunorubicin, intravenous, D₁-D₃, and 100 mg/m² Inj. cytarabine, intravenous, continuous infusion D₁-D₇, (3 + 7 protocol). The pre-sacral mass showed complete resolution following completion of treatment and bone marrow study was still normal. He is currently under strict surveillance with monthly follow-up and monitoring of complete blood count for the last one year, and is yet to develop any feature of AML.

DISCUSSION

Myeloid sarcoma is generally observed as a complication of AML, myelodysplastic syndromes, or myeloproliferative disorders, and it may be perceived as a *de novo* tumour without marrow involvement or as a tumour associated with leukaemia in the marrow, and as a site of leukaemia relapse.^[4-7] Myeloid sarcoma has been associated with 3.1-9.1% of AML cases.^[8] Rarely they can occur alone without peripheral blood or bone marrow evidence of leukaemia, thus representing a diagnostic challenge in patients without evidence of systemic disease. Late development of AML frequently occurs and the interval between initial diagnosis of myeloid sarcoma and onset of acute leukaemia has been reported as between 1 and 49 months (average: 10 months).^[9] All levels of the spine may be affected by myeloid sarcoma. The thoracic spine is most commonly involved (73%) followed by the lumbar (34%), sacral (23%), and cervical (5%) regions. Multiple spinal lesions have been diagnosed in only 18% of patients.^[9] However, myeloid sarcoma without bone marrow involvement is rare and only a few cases have been reported with spinal involvement.^[10] The origin of the tumour is thought to be bone marrow, with migration to extra-osseous locations through Haversian canals.^[9] Macroscopically it is usually green in appearance due to the presence of myeloperoxidase. Diagnosis is often difficult when myeloblastic cells are poorly differentiated and common misdiagnosis includes large cell lymphoma. Our patient was reported as a case of a small blue, round cell tumour by histopathology and diagnosis was apparent only after immunohistochemistry report was available.

The optimal treatment for isolated myeloid sarcoma without evidence of systemic disease is yet to be defined due to the rarity of this entity. Treatment options for myeloid sarcoma involving spine include local EBRT, decompression surgery, systemic chemotherapy, or any combination of these. Myeloid sarcoma is radiosensitive and the radiation dose required is also quite low, so local EBRT is often employed in patients with compressive myelopathy or radiculopathy. Neurological recovery is expected if radiation therapy is

started early in the course of the disease. Radiotherapy is also used for consolidation when induction chemotherapy results in partial remission of the tumour mass.^[11] Systemic chemotherapy is usually necessary in patients without evidence of systemic disease as radiotherapy alone fails to achieve cure for even such localised disease.^[9]

CONCLUSION

We report a case of a 22-year-old young male who presented with radicular pain of lower limbs with urinary retention and erectile dysfunction due to cauda equina compression by a pre-sacral mass, which was shown to be an extramedullary myeloid sarcoma by histopathology and immunohistochemistry. However, the patient did not have any evidence of AML either in peripheral blood or bone marrow. He responded favourably to treatment with local EBRT and systemic chemotherapy using the 3 + 7 protocol. So, early recognition of this rare entity is of paramount importance as prompt, aggressive, combined-modality management can achieve durable remission and preserve reasonable quality of life.

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