Pathological dislocation of the dorsal spine following granulocytic sarcoma in a non-leukaemic patient

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Abstract We describe a previously healthy, non-leukaemic young male presenting with neurological deficit and a pathological dislocation of D8 over D9 vertebra. The magnetic resonance imaging showed an enhancing soft tissue tumour. His basic laboratory workup as well as a bone marrow biopsy was normal. Through a single midline posterior approach, he underwent a decompressive laminectomy of D8 and D9 vertebra, anterior column reconstruction with a meshed titanium cage and posterior pedicle screw instrumentation. The histological diagnosis of granulocytic sarcoma was confirmed by appropriate immuno-histochemical studies. He received postoperative radiotherapy following which his wound dehisced and the tumour fungated and spread to his left thigh. He declined chemotherapy and unfortunately expired 9 months later. This case is presented to draw attention to the unusual presentation and to stress that granulocytic sarcoma should be kept in mind when making the differential diagnosis in patients with signs of spinal cord compression even in non-leukaemic individuals.

Keywords Granulocytic sarcoma · Non-leukaemia · Pathological dislocation · Surgery

Introduction

Granulocytic sarcoma is a tumour that originates from malignant myeloid blast cells [1, 2]. It rarely presents with a neurological deficit as the initial presentation in a non-leukaemic patient [3, 4]. Epidural granulocytic sarcoma may manifest variously as an acute spinal cord compression [5], triplegia [6], cauda equina syndrome [5], a radiculopathy [1] or sciatica [7]. We discussed the diagnosis and management of this condition in a young male who presented with spinal cord compression and a pathological dislocation of dorsal spine. To the best of our knowledge, this is the first report of a patient presenting in this manner.

Case report

A 15-year-old boy presented to us with mid-back pain for duration of 30 days and weakness in both lower limbs for 3 days. He had associated loss of weight and appetite. Prior to admission, he developed weakness in both lower limbs as a consequence of which he had a trivial fall in his bathroom after which the back pain had markedly increased. Bladder and bowel functions were normal. Local examination of the dorsal spine revealed a knuckle deformity at D9–10 vertebrae. Motor power was graded as Frankel D, bilateral ankle clonus with a positive Babinski sign was present. Sphincteric tone was normal. Apart from a mild anaemia (haemoglobin = 10.5% g), the other haematological parameters were unremarkable (white count = 9,200/mm³). The X-ray revealed a destruction of D9 with an anterolisthesis of D8 relative to D9 (Fig. 1). The magnetic resonance image showed lobulated soft tissue replacement of the posterior elements and posterior body of D8 and D9 vertebra, altered signal intensity and collapse of

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Fig. 1 Preoperative X-ray (lateral view) showing destruction of D9 vertebra and pathological dislocation at D8 over D9

D9 vertebra with a soft tissue epidural component extending from D8 to D10 (Fig. 2a, b). Through a single posterior approach, he underwent realignment and posterior stabilisation with pedicle screws, decompressive laminectomy of D8 and D9 vertebra, subtotal corpectomy of D9 vertebra and reconstruction of the anterior column with a titanium cage (Fig. 3).

Intraoperative pathological assessment by frozen section was suggestive of a malignant round cell tumour (lymphoma or Ewing's sarcoma/primitive neuroectodermal tumour were considered). Histological examination of the surgical specimen showed diffuse monotonous infiltrates of round, medium to large sized cells with scanty eosinophilic cytoplasm and round to folded nuclei, coarse chromatin and inconspicuous nucleoli (Fig. 4). A few interspersed cells with notched nuclei were also seen. On immunohistochemistry, these tumour cells showed cytoplasmic positivity for myeloperoxidase (Fig. 5) and membranous staining for CD43 (Fig. 6) and CD99. Neoplastic cells were negative for CD3, CD20, CD30, Desmin and Myf-4. The morphological and immuno-histochemical features were consistent with the diagnosis of granulocytic sarcoma. A bone marrow examination revealed a mildly hypercellular marrow with no evidence of malignant changes. He was ambulated on the third postoperative day and reported relief of pain. His neurological status improved to Frankel E in about 6 weeks when he was independently ambulant. He was advised postoperative chemotherapy, but declined and as an alternative was prepared for postoperative radiotherapy (30

Fig. 2 **a** Preoperative T2 weighted sagittal. Magnetic resonance image showing altered signal intensity and collapse at D9 vertebra, anterolisthesis of D8 over D9 with a soft tissue epidural component extending from D8 to D10. **b** Preoperative T2 weighted axial magnetic resonance image showing the predominantly posterior intraspinal soft tissue lesion with destruction of the facet joint and posterior elements

Fig. 3 Postoperative X-ray (lateral view) showing corpectomy of D9 vertebra, realignment and reconstruction of the anterior column with a titanium cage and adjuvant posterior stabilisation with pedicle screws from D6, D7, D10 and D11 vertebra

Grey in 10 cycles) 2 weeks after removal of sutures. Unfortunately after two cycles of radiotherapy, the surgical wound broke down and the tumour rapidly fungated and

Fig. 4 Sheet of medium to large sized cells with a thin rim of eosinophilic cytoplasm and round to convoluted dark staining nuclei (hematoxylin and eosin stain, $\times 400$)

Fig. 5 Cytoplasmic positive immunostaining in tumour cells for myeloperoxidase ($\times 400$)

spread to appear as a skin lesion in the left thigh. At follow-up of 5 months the patient had a normochromic normocytic blood picture, total white count = $22,000/\text{mm}^3$, neutrophil = 86%, lymphocyte = 8%, monocyte = 6%, no blast formation. After this the patient did not follow up and we learnt of his death through a telephonic interview with his parents 9 months later.

Discussion

Granulocytic sarcoma is a localised mass of primitive myeloid cells of the granulocytic series that has infiltrated an extramedullary site [8]. They are usually seen in association with acute myeloblastic leukaemia (AML). Rarely they may occur in otherwise healthy patients without any

Fig. 6 Positive membranous immunostaining in tumour cells for CD43 ($\times 400$)

evidence of systemic disease and this may then herald the appearance of leukaemia often within weeks to months [5, 8] at an average of 10.5 months [6].

Granulocytic sarcoma usually occurs in bone, skin and lymph nodes [5, 6]. Bony involvement is most common in the orbit, sacrum, ribs and spine [6]. The origin of the tumour is thought to be in the bone marrow with migration to extra osseous locations achieved via the Haversian canal [5, 8]. When the cut surfaces of the specimen show a green colour because of myeloperoxidase enzyme within the cells, these tumours are called chloroma (from the Greek *klóros*, green). As they are solid tumours, granulocytic sarcomas originating in the spinal column may cause cord compression and present acutely as neurological deficits.

Though a rare occurrence, pathological dislocation of the dorsal spine has been described in tuberculosis [9] Charcot's disease [10] and neurofibromatosis [11] and occurs when the disease process destroys or weakens the bony restraint between two adjacent vertebra predisposing them to dislocate. This is to the best of our knowledge, the first instance when a pathological dislocation has been caused by granulocytic sarcoma.

Without the clue of circulating myeloblasts and the absence of the characteristic green colour, the appearance of a granulocytic sarcoma in a non-leukaemic patient presents a diagnostic challenge [1, 12]. In these patients, the histological differential diagnosis of granulocytic sarcoma includes malignant lymphoma [13], Ewing's sarcoma/primitive neuroectodermal tumour (PNET), Langerhans histiocytosis X and metastatic tumours [1]. Granulocytic sarcoma consists of histologically uniform immature cells, but often includes mature neutrophils and immature eosinophils. Therefore, immuno-histochemistry is effective and should be performed to confirm the diagnosis of GS. Myeloperoxidase, CD 15 (granulocyte-associate antigen)

and neutrophil elastase are especially effective to differentiate granulocytic sarcoma from other tumours. CD 99(MIC2), synaptophysin and neurofilament protein are positive for Ewing's sarcoma/PNET, but negative for granulocytic sarcoma. CD3, CD 56 and CD 79 α are useful for differentiating GS from malignant lymphoma. CD 1a and S-100 protein are positive for Langerhans histiocytosis X and cytokeratin for metastatic tumour, but all are negative for granulocytic sarcoma [1].

Treatment options for patients presenting with spinal granulocytic sarcoma include, radiation therapy, chemotherapy, bone marrow transplantation [1, 5], surgical decompression [8] or a combination of these treatment methods. The treatment strategy must account for not only the localised tumour but also the concurrent (or imminent) leukaemia [8]. Though granulocytic sarcoma is radiosensitive, it must be used cautiously in patients who have been operated as there is the hazard of wound breakdown as was seen in this patient. In view of the potential transformation to AML, prophylactic systemic chemotherapy is used in newly diagnosed cases [6, 8]. Surgical decompression is warranted in patients who present with a neurological deficit. In the present case, the patient had features of dorsal cord compression along with a mechanically unstable spine due to a pathological dislocation. Our surgical goals were canal decompression, tumour clearance, vertebral column alignment stabilisation and reconstruction to permit early ambulation. Through a posterior approach, we performed a laminectomy of D9, excision of D9 vertebra, anterior column reconstruction with a Titanium mesh cage filled with bone cement between D8 and D10 and then stabilisation of the posterior column with pedicle screws. The patient reported substantial pain relief, his early neurological deficit recovered, and he was able to ambulate independently within 4 weeks of the surgery. In the follow-up X-rays (Fig. 3) the cage appears to have marginally encroached into the spinal canal (this may be due to settling of the cage). However, as the patient had a satisfactory neurological recovery in the postoperative period and was ambulant without support we did not consider a postoperative MRI to assess the dural sac.

This may be the first instance when a patient with GS has been managed with this surgical strategy.

In conclusion, we stress that granulocytic sarcoma should be considered when making the differential

diagnosis in patients with signs of spinal cord compression. An early diagnosis is mandatory. Surgical intervention tailored to the patient's presentation followed up with appropriate chemotherapy and radiotherapy is necessary. The potential of wound breakdown after radiotherapy must be borne in mind.

Conflict of interest statement None of the authors has any potential conflict of interest.

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