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Epidural spinal cord compression as initial clinical presentation of an acute myeloid leukaemia: case report and literature review

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Abstract

Epidural localization of myeloid leukaemia is rarely reported. Spinal cord compression as an initial presentation of acute myeloid leukaemia is extremely rare. This is a report of a 17-year-old black boy who presented to emergency department with neurological symptoms of spinal cord compression. Imaging modalities showed multiple soft tissue masses in the epidural space. After surgical treatment, histopathological examination of the epidural mass showed myeloid leukaemia cells infiltration. Literature review on Medline and "scholar Google" database was done. The characteristics and management of extra-medullary leukaemia are discussed. Granulocytic sarcoma, myeloid sarcoma or chloroma with acute myeloid leukaemia should be considered as part of epidural spinal cord compression. Therefore surgery is indicated on an emergent basis.

Keywords: Spinal cord compression, Initial epidural localization, Acute myeloid leukaemia

Background

Acute myeloid leukaemia corresponds to malignant monoclonal proliferation of medullary blastic myeloid cells with an interrupted differentiation [1]. Clinically acute myeloid leukaemia will result in adenopathy and hepatosplenic syndrome associated with bone marrow failure syndrome. Extra nodal involvement is rare and mostly made of frequently observed forms of lymphoblastic leukemia in children. These attacks represent 2 to 8 % of acute myeloid leukaemia. Epidural localization is unusual and is a rare cause of spinal cord compression. Spinal cord compression as an initial presentation of acute myeloid leukaemia is extremely rare. In this study, we did not consider Spinal cord compression as the initial presentation of acute biphenotypic leukaemia [2]. The authors reported a case of acute myeloid leukaemia revealed by thoracic spinal cord compression. Literature review follows the case report.

Clinical case

A 17-year –old A black patient with no known significant medical history, complained of a 3-week history of inter-scapular back pain progressively exacerbating with insomnia associated with paraesthesia, hypoesthesia of the lower limbs and trunk. The rapidly worsening deficit led to emergent Neurosurgery consultation. Neurological examination found an almost paraplegic patient with increased deep tendon reflexes, a bilateral Babinski, a distended bladder, hypoesthesia and dysesthesia with at T10 sensitive level. Further clinical examination found no evidence of lymphadenopathy or hepatosplenomegaly. The diagnosis of spinal cord compression was made and a spinal CT scan showed an extensive epidural mass at T4-T9. MRI confirmed the presence of a thoracic epidural mass posterior to the spinal cord between T4 and T9 (Fig. 1a, b, c, d, e, f). The MR imaging showed hypointense lesion on T1 weighted sequence, isointense on T2 weighted sequence and enhanced on T1 with gadolinium. There was a distinct line of dura demarcation between the tumor mass and the spinal cord.

Full blood count revealed leucocytosis with a importantly increased white blood cell count 200,000/mm^3, with 77 % blasts, haemoglobin: 10.7 g/$dL^{-1;}$ platelet count: 102 × 10^9 L^{-1}. The myelogram diagnosis was acute myelogenous leukaemia. Histologically the tumor is composed of immature cells. There are some immature eosinophile and neutrophile. Cytogenetic evaluation

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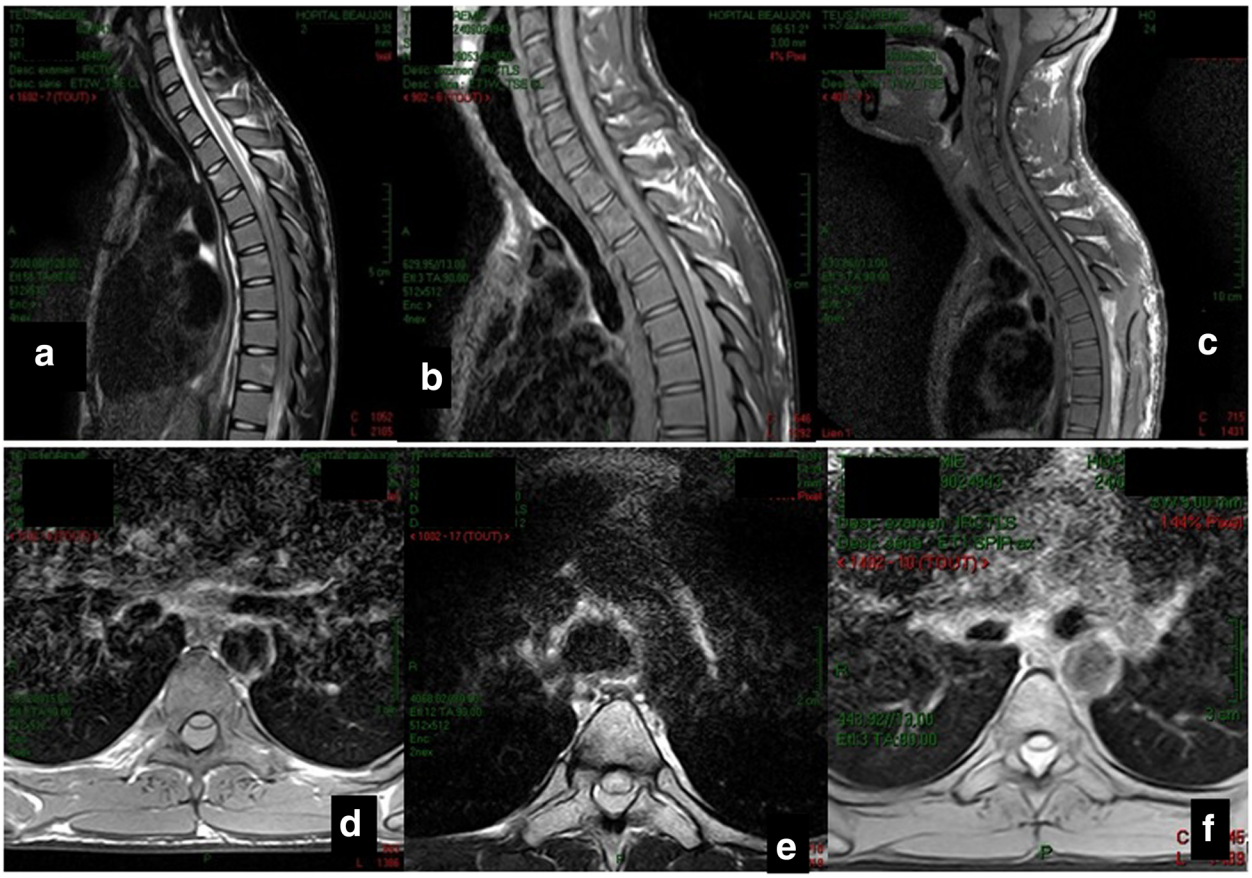

Fig. 1 Spine MRI showing spinal cord compression at T4-T9 level by posterior epidural mass. **a** Sagittal view showing a T4T9 posterior mass which is isointense on T2 weighted sequence. **b** Sagittal view T1 sequence showing a hypointense T4T9 posterior lésion well demarcated from the dura. **c** Sagittal view showing a contrast enhancement of the T4-T9 lesion. **d** Axial view: The lesion is hypointense on T1 weighted sequence. **e** Axial view: The lesion is isointense on T2 sequence at the level of T5. **f** Axial view showing the enhancing lesion at the level of T5 wth a good demarcation from the dura

could not be done. Additional blood test did not find bleeding disorders or ion anomaly. Given the severity of the neurological picture and predicted rapid deterioration, a laminectomy from T4 to T10 was performed in emergency and found that the posterior epidural space was occupied by greyish, haemorrhagic tumour tissue, completely separated from the dura and pushes forwards. A macroscopically complete tumour resection was performed. Haemostasis was difficult. Histopathological examination of the tumour mass was favourable to leukemic blast cells. It was type 2 leukemia (Fig. 2). Advanced treatment of acute

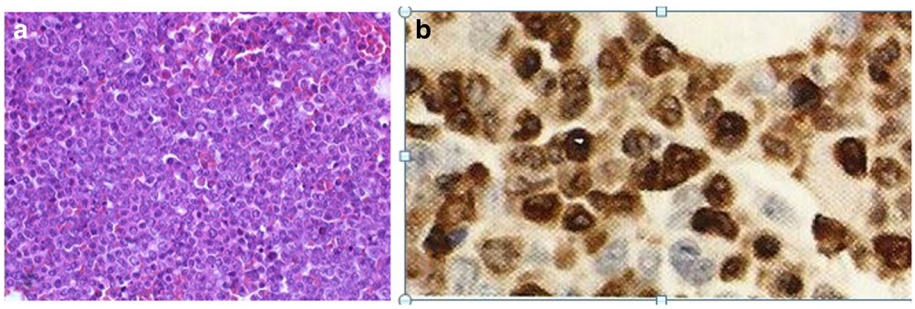

Fig. 2 HE X 100: Tumor proliferation with blasts exhibiting cytonuclear atypia (**a**). Immunohistochemistry X 200: Nuclear expression of antibodies anti myeloperoxidase (MPO) by the majority of undifferentiated blasts (**b**)

Table 1 Literature review from 2005 to 2015. characterization of acute myeloid leukemia associated to spinal cord compression

Nb	References	Sex/age (years)	Cytogenetic	Histology and immunohistochemistry	Localization	signs	MR Imaging	Laboratory findings	Delay and diagnosis treatement and outcome
1	Meltzer et al. 2005 [3]	M/10	-	-Myeloblastic Acute myeloid leukemia-Chloroma	T2-T6	-Midback pain-weight loss-decreased sensation to vibration and light touch over the left leg -decreased sensation to pain and temperature in the right leg	-paravertebral lesions with spinal-posterior epidural and prevertebral mass	WBC 12,800/mm^3, HB 9.4 g/dl, platelet 74,800/mm^3.-lactate dehydrogenase was 570 IU/l,-immature cells	-3 weeks-Laminectomie + Chemotherapy + spinal radiation-Total Neurological improvement after 6 month-well more than 18 months of therapy
2	Ghalaut et al. 2005. [4]	23/M	-	Granulocytic sarcoma due to acute myeloid leukemia	T4-T6	-Backache-numbness of bilateral legs-sensory loss up to mid thoracic region anemia	-Epidural mass extending from T4-T6 with marrow signal alteration involving T1 and T5 to T7	-	- surgical excision + chemotherapy -neurological improvement
3	Amalraj et al. 2009 [5]	14/M	-	Granulocytic sarcoma promyelocytic leukemia AML M3	T3-T10	-Acute paraplegia -weakness of both lower limbs-numbness of both lower limbs-back pain for the last-bladder symptoms	hypointense in T1 hyperintense in T2 dorsally compressing the spinal cord	Hemoglobin 13 gm%, total count 8,400, polymorph 60 %, and lymph 40 %, with high erythrocyte. RBC mild hypochromic microcytic, WBC count normal predominantly blast cells with high nuclear cytoplasmic ratio, moderate cytoplasm, hyperchromatic nuclei with multiple nucleoli, platelet count decreased.	-paraplegia improved with radiotherapy and chemotherapy
4	Olcay et al. 2009 [6]	M/12	Cytogenetic evaluation could not be done.	acute myeloblastic leukemia CD45:95.15 %, CD15: 35.56 %, CD33:77.57 %, CD117:79.3 %, CD34:43.38 %, CD13:29.32 %. HLA-DR:86, 4 %, CDw65:23.9 %, CD19:49.54 % CD20:21.65 %, CD14:3.14 %.	Lumbar (L3) -Conus medullaris-left maxillary mass	-Conus medullaris syndrome	Cauda equine nerve roots were thickened and clumped in the left anterolateral side of the spinal canal.	Hb:8 g/dL WBC : 7.4 × 109/mm^3 Thrombocyte:60.10^9/mm^3 Lactate dehydrogenase: 691 U/l	-Chemotherapy + triple intratecal therapy-Radiotherapy-Symptom partially resolved died because of pneumonia and disseminated intravascular coagulation
5	Yin et al. 2010 [7]	28/M	-	myeloid sarcoma with acute myeloid leukemia CD68, CD45, CD43, CD117 and lysozyme but not of MPO, CD20	T12-L1	-low back pain-numbness of his legs-bladder-incontinence-hypoesthesia below T12 paraparesis	-Posterior epidural mass T12-L1	White blood cell 3,900/mm^3- hemoglobin :11.0 g/dL-hematocrit of 38.2 %.	High-dose methylprednisolone -Decompression with T12-L1 laminectomy and tumor resection. Paraparesis Improvement - anti-AML therapy, but died of sepsis
7	Bittencourt et al. 2011 [8]	53/M	t(15;17)(q22q21) abnormality	-Acute promyelocytic leukemia presenting as extradural mass-PCR	T6-T8	-Progressive fatigue, pain legs weakness Paraparesis- Gingival	Posterolateral extradural mass from	Hb:10,6 g/dl^{-1} WBC: 7,5.10^9/L^{-1}	delais4 months/ chemotherapy (daunorubicin) +

Table 1 Literature review from 2005 to 2015. characterization of acute myeloid leukemia associated to spinal cord compression *(Continued)*

				for the PML-RARα gene was positive		hemorrhage, hepatomegaly	T6-T8 with medullar compression	Platelet:12.10^9/l-1 Fibrinogen:1.4 g/L-^{1} INR: 1.86 (N:0,8-1,2) Uric acid and lactate dehydrogenase were both elevated	radiotherapy/Evolution: no neurologic improvement + died of sepsis
8	Kyaw et al. 2012 [9]	26/M	-	-Myeloid sarcoma: acute promyelocytic leukemia-CD33, CD117, CD64, CD34 and cytoplasmic MPO were presented Reverse transcriptase-polymerase chain reaction showed BCR1-type PML-RARα fusion copies	T2-T4 and T12 – L2	Progressive back pain and bilateral leg weakness: -paraparesis Loss of pain and sensory perception	Multiple hyperintense T12,L1,L2, L4 and L5 vertebral bodies and sacrum Intraspinal extradural masses L4 and L5 vertebral bodies and sacrum Intraspinal extradural masses located fromT2 toT4 and T12 to L2.	Hb:10,7 g/dL^{-1} WBC:2,8.10^9/L^{-1} Platelet : 102.10^9/L^{-1}	-Radiotherapy-Chemotherapy: retinoic acid + ida rubicin-Neurological improvement - Good remission
9	Ben et al. 2013 [10]	21/M	karyotype: 46, XY, t (8, 21) (q22; q22) / 46, XY	chloromaCD3, CD20 = --positive for myeloperoxidaseacute myeloblastic leukaemia (AML) of the French-American-British M2 subtype	T4/ T7, T1-T2	Progressive paraplegia. urinary retention	intermediate in T1, hyperintense in T2	hemoglobin 8 g/dL^{-1}, white blood cell count 3100/mm^3, platelet count 44000/mm^3 and 3 % neutrophils, 44 % lymphocytes and 44 % .	laminectomy + tumour was totally removed -rubidomycine (45 mg/m^2 daily for 3 days) and cytosine arabinoside (200 mg/m^2 continuous infusion for 7 days). -improvement paraplegia- complete remission
10	Krishnamurthy et al. 2014 [11]	16 /M	-	Granulocytic sarcoma associated with leukaemia,-acute myeloblastic leukaemia (AML) of the French-American-British M2 subtype.-subleukemic leukemia		Low backaches follow proptosis, spinal cord causing significant compression of the spinal cord.	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	Hb : 7.2 g/dl, 3 RBC 2.5 million/mm^3,TC : 11200 cells/mm^3, with a differential ount of N 41 %, L-28 %, E-01 %, M-30 %, 3ESR 120 mm/hr, Platelet count: 51,000/mm^3, PT/INR –- dimorphic anemia, thrombocytopenia and myeloblasts	-radiotherapy and chemotherapy
11	Present case.	17/M	-Cytogenetic evaluation could not be done.	-Myeloid leukemia-type 2 leukemia	T4T9	Acute paraplegia	hypointense lesion on T1 weighted sequence isointense T2 weighted sequence enhanced on T1 with gadolinium. (epidural mass (hyperintense)	blood count revealed leucocytosis with a major white blood cell count 200,000 / mm^3, with 77 % blasts, hemoglobin: 10.9 g dl; platelet count: 106 × 10^9 L^{-1}	-Chemotherapy-Died one week after diagnosis

Literature review from 2005 to 2015. characterization of acute myeloid leukemia associated to spinal cord compression
N b: Number

myeloid leukaemia was indicated. But the patient died a week after the diagnosis of spinal cord compression because of sepsis.

Discussion

The spinal cord compression revealing acute myeloid leukaemia is unusual. Nine cases have been reported from 2005 to 2015 [3–11] (Table 1). It often appears like a solid tumour known as granulocytic sarcoma or myeloid, or also chloroma as seen in our patient [12]. It corresponds to the migration outside of the bone marrow of myeloid cells that proliferate on their own [1]. The first case was described in 1811 by Burns in 1893 and Dock who reported its association with leukaemia [13, 14]. It is reported in 3.1 to 9.1 % of patients with acute myeloid leukaemia [15]. We found ten cases from 2005 to 2015 which were reported in literature. In those cases epidural space was occupied by granulocyt sarcoma [3-9 11, 15]. Surgically excision of the tumour appeared to be the first treatment option in these cases. Granulocytic sarcoma is frequently diagnosed simultaneously with or after the start of an acute myeloid leukaemia or may be the initial sign of a relapse in a patient in remission [3–9, 11, 15]. In non-leukemic patients, myeloid granulocytic sarcoma usually precedes acute myeloid leukaemia [16]. In 87 to 88 % of patients without hematologic abnormalities at diagnosis, acute myeloid leukaemia develops in the 10.5 to 11 following months [17]. Myeloid sarcoma may occur in all tissues but they are frequently localized on the skin, bone, soft tissues of the head and neck (especially on the orbit) and adenopathies [18]. Spinal localization and especially epidural is rare. Thoracic localization is the most frequent spinal localization followed by lumbar and sacral localizations. Clinical signs are in most cases made of spinal cord compression. Sometimes they involve isolated rachialgia [3–9, 11, 15]. MRI is the best choice for neuroimaging examination to show epidural tumour lesions. It is also the best neuroimaging examination [3–9, 11, 15] to show the epidural tumour lesions without being specific. Granulocytic sarcoma is isointense in T1 and T2 with in general a contrast enhancement [3–9, 11, 15]. The therapeutic strategy of this myeloid sarcoma is based on chemotherapy, radiation therapy, surgical decompression and bone marrow transplantation and any combination of these methods [3–9, 11, 15]. 1n cases of granulocytic sarcoma in particular by acute myeloid leukaemia with neurological signs, priority should be given to chemotherapy and/or radiation therapy rather than surgery. In our patient the diagnosis of acute myeloid leukaemia was made from full blood count after surgery. Surgery was only justified by the acuteness and quickly worsening neurological disorders. Despite some cases of more or less complete neurological recovery with a variable remission, most patients present with neurological sequellae and die from haematological complications dominated by sepsis.

Conclusion

This case associated with the literature review has shown that granulocytic sarcoma with acute myeloid leukaemia should be considered as part of epidural spinal cord compression .it might be the initial sign of epidural spinal cord compression . Therefore surgery is indicated in emergency. Chemotherapy and radiotherapy are secondly introduced according to the diagnosis.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

DNO has performed the surgery, evaluated the patient, and drafted and revised the manuscript. ABB assisted the surgeon at the operation room, grammatically reviewed the manuscript and participated to the discussion. AVT evaluated the patient, carried out the literature review. LD evaluated the patient, carried out the literature review. All authors read and approved the final manuscript.

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