EMERGENCY CASEBOOK

Acute myeloid leukaemia presenting as faecal incontinence

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Epidural sacral nerve compression as an initial feature of leukaemia is a rare complication. The findings in a 16-year-old boy who presented to an emergency department with symptoms of faecal incontinence are reported herein. Radiological imaging demonstrated soft-tissue masses in the sacral epidural space. The diagnosis of acute myeloid leukaemia was confirmed on bone marrow aspirate. The characteristics and management of extramedullary leukaemia are discussed.

A 16-year-old boy developed faecal incontinence and sacral pain 3 days before his presentation to the emergency department. He denied having trauma, fever, night sweat and leg pain. Medical history was not significant, except for otitis media 1 month previously. The symptoms of otitis media had remained.

On physical examination, he had normal vital signs, anicteric sclera, pale conjunctiva and did not have palpable lymph nodes. His chest was clear to auscultation. Examinations of the heart, abdomen and skin did not show abnormalities. There was no scoliosis, kyphosis, or vertebral or costovertebral tenderness. Cranial nerves and motor function were intact. But there was hypoaesthesia in S3 and S4 perianal areas, and tenderness in the coccygeal and sacral areas. Anal sphincter tone was normal. He was able to walk. The remainder of his neurological examination was normal.

Laboratory findings showed a haemoglobin level of 5.8 g/dl, a platelet count of 30 000/mm³ and a white blood cell count of 208 000/mm³, with 10% neutrophils, 2% lymphocytes, 0% monocyte and 52% blastocytes. The serum lactate dehydrogenase was 2615 IU/l (reference range 219–480 IU/l). The patient received intravenous dexamethasone in the emergency department, and MRI of lumbar spine was performed immediately, as we suspected spinal cord compression. A T2-weighted imaging showed an epidural mass compressing the sacral nerve root from the first to the second vertebral body of the sacrum (fig 1A), while the axial section showed that the sacral nerve root was pressed and deformed by the tumour (fig 1B). The mass had lower signal intensity in T1-weighted imaging and higher signal intensity in T2-weighted imaging than the surrounding muscles. The findings suggested lymphoma or leukaemic infiltration. A bone marrow aspiration and biopsy were performed, which established the diagnosis of acute myeloid leukaemia (AML). Cerebrospinal fluid analysis showed no malignant cell.

The patient was treated with intrathecal and induction chemotherapy. His condition improved on the third day following admission, with improvement of faecal incontinence and pain in the sacral area. Re-evaluation in hospital on day 21 showed complete disappearance of the sacral epidural mass.

DISCUSSION

Epidural cord compression is an uncommon initial presentation of leukaemia. It is described more frequently in AML, followed

by chronic myeloid leukaemia and lymphoma.^{1 2} Other cases of spinal cord compression secondary to leukaemia have been reported.^{2 3} Most patients develop back pain, abdominal pain, or pain in the lower extremities or perianal region.³ Muscle weakness, ranging from lower extremity weakness to complete paraplegia, is another common symptom. As in all cases of cord compression, urinary retention and incontinence are ominous signs. The thoracic cord is considered the most common site, followed by lumbar levels. This patient differs from most other cases by virtue of a sacral epidural mass, in addition to the atypical presentation of AML. His neurological symptoms were only faecal incontinence and hypoaesthesia in the perianal area. Sacral nerve root compression by sacral epidural mass is a very rare initial presentation of leukaemia.

The differential diagnosis of spinal cord compression includes a haematoma, abscess, meningioma, metastasis, lipomatosis and lymphoma. These extramedullary collections of leukaemia cells are usually chloroma. These leukaemia cell masses usually expand rather slowly, which is probably a result of a suboptimal growth environment. As in our patient, the initial suspicion of leukaemia may not be easy when the initial signs and symptoms result only from chloroma.

Treatment consists of local irradiation and high-dose corticosteroids along with systemic chemotherapy with or without laminectomy. Considerable controversy exists because there are no convincing data to support the use of one treatment over another. This is largely because spinal cord compression secondary to leukaemia is uncommon. But irreversible neurological damage is always a concern in patients with cord compression. Steroids are routinely administered as emergent therapy. Dexamethasone is advised for patients with spinal cord compression caused by malignancy, because of its direct oncolytic effect and its effect in decreasing the water content of the spinal cord.⁴

Laminectomy does not appear to be necessary in patients with newly diagnosed leukaemia and lymphoma. It should be limited to biopsy in case diagnosis is doubtful. As the optimal treatment of spinal cord compression with AML has not been established, we employed initial intrathecal and induction chemotherapy with high-dose dexamethasone and reserved local irradiation in case of further deterioration. Our patient recovered fully without neurological sequela. We conclude that patients with atypical neurological symptoms such as hypoaesthesia in the perianal area and faecal incontinence should be closely observed, and that the emergency doctors should suspect an epidural cord compression and make every effort to diagnose it. Neurological function can be preserved in most of these patients if the diagnosis is made early and the treatment is undertaken before irreparable damage occurs.

Abbreviation: AML, acute myeloid leukaemia

Figure 1 Spinal MRI without contrast. (A) T2-weighted sagittal image showing an epidural mass compressing the sacral nerve root from the first to the second vertebral body of sacrum. (B) T2-weighted axial image showing the sacral nerve root compressed and deformed by the tumour.

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