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Sickle cell disease of the spine in children

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Objective: To determine the incidence of back pain in children admitted with sickle cell disease so as to increase awareness of this disease in black children presenting with back pain or discomfort.

Design: A retrospective review.

Setting: The Children's Hospital of Eastern Ontario in Ottawa, a tertiary care, university affiliated centre.

Patients: Thirty children were treated for active sickle cell disease between 1990 and 1996. Eleven (5 boys, 6 girls) suffered vascular-occlusive phenomena occurring in bone, referred to as "bony crises," requiring a total of 49 admissions.

Main outcome measures: Clinical manifestations of spinal involvement by sickle cell disease.

Main results: The spine represented the second most common area of bone involvement (26%) exceeded only by the knee (35%). The vertebral level affected was lumbosacral in 66% of cases, followed by thoracic in 22% and cervical in only 12%. Eighty-six percent of the children with spinal pain were anemic upon presentation, 71% had an elevated leukocyte count, 15% were hyponatremic and 15% were hyperkalemic. Minimal physical signs in the spine were noted, other than a local tenderness over the spinous process in 71% and a decreased range of back motion in 17%.

Conclusions: Sickle cell disease is becoming more common in Canada as a result of increasing immigration from African countries and should always be considered as a possible cause of back pain in a black child.

Objectif : Déterminer l'incidence de la dorsalgie chez les enfants hospitalisés atteints de dépranocytose, afin de faire connaître davantage cette maladie chez les enfants de race noire qui ont des douleurs ou des malaises au dos.

Conception : Étude rétrospective.

Contexte : Hôpital pour enfants de l'est de l'Ontario à Ottawa, hôpital de soins tertiaires affilié à une université.

Patients : Trente enfants ont été traités pour une dépranocytose active entre 1990 et 1996. Onze d'entre eux (cinq garçons, six filles) présentaient une occlusion vasculaire dans les os appelée «crise osseuse» qui a entraîné au total 49 admissions.

Principales mesures de résultats : Manifestations cliniques d'atteinte de la colonne par la dépranocytose.

Principaux résultats : La colonne est le deuxième endroit où l'atteinte osseuse est la plus fréquente (26 %), après le genou (35 %). Le niveau vertébral atteint était le niveau lombo-sacré dans 66 % des cas, thoracique dans 22 % des cas et cervical dans 12 % des cas. Quatre-vingt-six pour cent des enfants qui avaient des douleurs à la colonne étaient anémiques lorsqu'ils se sont présentés, 71 % avaient une numération leucocytaire élevée, 15 % étaient hyponatrémiques et 15 % étaient hyperkaliémiques. On a constaté des signes physiques minimes à la colonne, à part une sensibilité locale au niveau de l'apophyse épineuse dans 71 % des cas et une diminution de l'amplitude du mouvement du dos dans 17 % des cas.

Conclusions : La dépranocytose se répand au Canada en raison de l'immigration croissante en provenance de pays africains et il faut toujours l'envisager comme cause possible de dorsalgie chez un enfant de race noire.

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Sickle cell disease is a well-known genetically determined hemolytic anemia that occurs almost exclusively in the black population. It is characterized by vascular occlusive episodes, visceral sequestration and aplastic or hemolytic crises. Vaso-occlusive crises result from intravascular sickling causing embolism with end-tissue microinfarction. These crises occur most commonly in bone, lung, spleen, brain and penis. Bony involvement has been reported to be most common in the lumbosacral spine, knee, shoulder, elbow and hip, and less commonly in the sternum, ribs, clavicle, calcaneus, iliac crest, mandible and zygoma.[1–4] Over the last few years we have seen an increasing number of black children presenting with recurrent back pain secondary to a sickle cell crisis.

The purpose of this study was to investigate the clinical manifestations of a Canadian pediatric population presenting with sickling crises involving the spine, in an attempt to increase the awareness of sickle cell disease as a possible cause of low back pain in the black child presenting with neck or back discomfort.

Table I

Distribution of Bony Crises in 30 Black Children Having Sickle Cell Disease Who Were Admitted to Hospital

Location	% of admissions
Knee	35
Spine	26
Elbow	10
Hip	10
Shoulder	7
Sternum	4
Ankle	3
Hand-foot	2
Wrist	2
Rib	1
Clavicle	0

Methods

We reviewed the charts of 30 black children, 13 boys and 17 girls, admitted with a diagnosis of sickle-cell-related bony crises to the Children's Hospital of Eastern Ontario, Ottawa, between 1990 and 1996 inclusively, focusing particularly on spinal involvement. There were no children with Mediterranean type sickle cell disease. The following information was obtained on each child: the sites of bony involvement, the affected spinal level, the laboratory findings, the clinical picture, the findings on physical examination, the radiologic features and the treatment.

Findings

Twenty-six percent of all bony crises that resulted in hospital admission partially or solely involved the spine. This was the second most common site of bony involvement, the first being the lower extremity (Table I). Of the 30 patients, 11 (5 boys, 6 girls) had a spinal complaints through the course of their disease. A total of 49 admissions were partially or solely related to a bony crisis involving the spine. No child presented with other spinal disease such as discitis or tumour.

The symptomatic spinal level was lumbosacral in 66% of the 49 admissions, thoracic in 22% and cervical in 12%. From the chart review, 86% of the cases were sufficiently well documented to determine the vertebral level.

Children with the most admissions for spinal bony crises also had the most marked radiologic spinal abnormalities. These usually were biconcave "codfish" or "H-shaped" vertebrae found most often in the lumbar spine but also in the thoracic spine (Fig. 1). Vertebral wedging associated with osteoporosis was also noted (Fig. 2), consistent with minor pathological compression fracture, which also would contribute to the back pain. Computed tomography was helpful in ruling out sepsis or tumour as a cause of vertebral compression in a few children with newly diagnosed sickle cell disease, but in general it did not add more information than that seen on plain radiographs.

The average hemoglobin level on admission was 82 g/L. The average leukocyte count and erythrocyte sedimentation rate were elevated at 18.0 $\times 10^9$/L and 36 mm/h respectively. The average serum sodium (138 mmol/L) and potassium (4.0 mmol/L) levels were normal, although 15% of the patients were hyponatremic and 15% were hyperkalemic. When considering the normal variations of hemoglobin with age, we found that 86% of children were anemic and 71% had a leukocytosis. On hemoglobin electrophoresis all children were found to be hemoglobin SS.

Just under half of the children with sickle cell crises affecting the spine had an elevated body temperature. Nausea was present in 22% and vomiting in 15% of the 49 admissions. Local tenderness was present over the posterior spine in 71% of the children, and a decreased range of spinal motion noted in 17%. Although some children were initially thought to have an infection, a negative blood culture, absence of a paraspinal abscess on imaging and the presence of hemoglobin SS on electrophoresis confirmed sickle cell disease as the cause of the severe back pain.

The standard therapy was rehydration at 1.67 times the maintenance level, administration of 35% oxygen and the use of blood transfusions, analgesics and antibiotics as needed. Two patients received treatment with hydroxyurea. The analgesic most commonly used to alleviate the back pain was acetaminophen (86% of the 49 admissions); however, the severity of the pain required morphine in 67%, codeine in 57% and demerol in 39% of cases.

Discussion

Sickle cell crisis is often associated with throbbing bone pain without any other physical findings. Anemia and leukocytosis are almost always present. Occasional local tenderness, erythema, warmth and swelling are present.

In a patient presenting with fever, leukocytosis and well-localized physical signs, it can be extremely difficult to distinguish microinfarction from osteomyelitis. Bone scanning does not help in the diagnosis since normal and increased uptake may be seen in both conditions.[5,6] Magnetic resonance imaging is more specific in qualifying the abnormality.[7] It is interesting that no infectious processes were found in our study, contrary to previous reports in adults with bony crises in sickle cell disease.[7,8] The most common organisms causing osteomyelitis in sickle cell disease are *Salmonella* (70%), *Staphylococcus* (15%) and *Streptococcus pneumoniae* (15%).[6,9] Our experience demonstrates that bone and spine pain in black children with hemoglobin SS is secondary to a bony crisis rather than sepsis. Aspiration of the involved long bone with a no. 16 trochar needle was helpful in determining that infection was not the etiology of the pain in several children. No trochar aspirations of spinal vertebrae were performed.

The skeletal changes are due to marrow hyperplasia and sickling, resulting in ischemia, infarction and occasional infection. The infarcts may occasionally form a nidus in which bacteria may thrive.[9] Further hypoxia and sickling may exacerbate the preexisting infarction. The marrow in the vertebral body continues to be erythropoietic and undergoes hyperplasia even in adult life, leading to trabecular destruction and marrow replacement by connective tissue.[10,11] As a result, the vertebrae are weaker and more likely to collapse with further aseptic necrosis and infection. Vertebrae often become flattened, with a

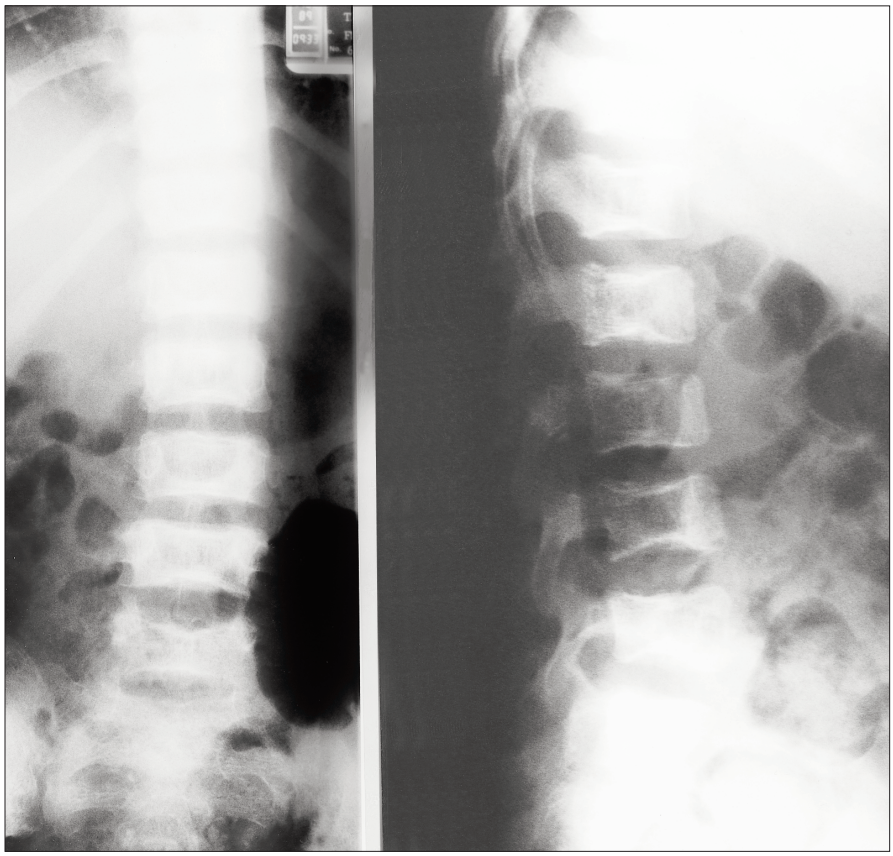

FIG. 1. Anteroposterior (left) and lateral (right) radiographs of the lumbar spine of a 15-year-old boy with sickle cell anemia, showing typical biconcave "H-shaped" or "codfish" vertebrae.

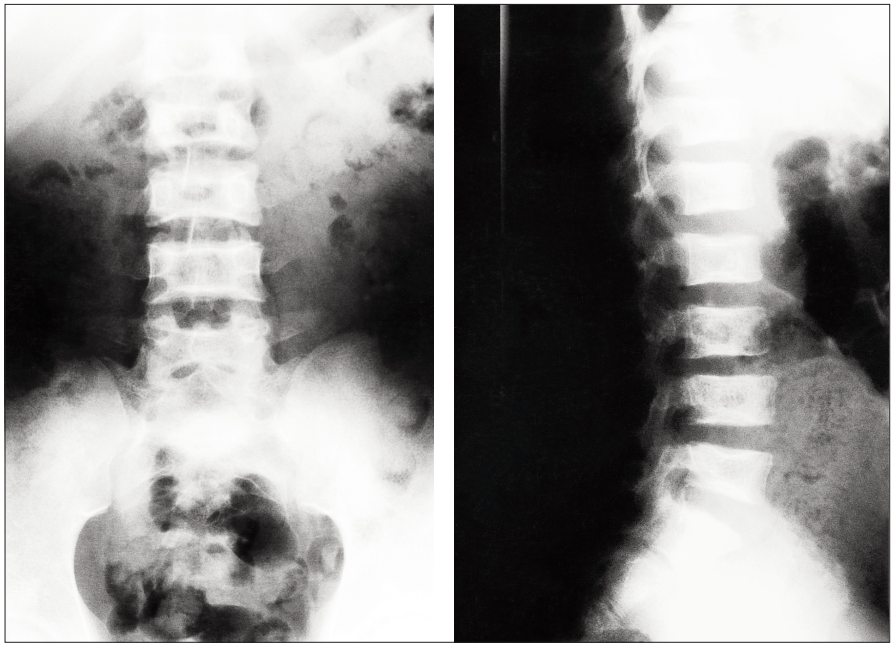

FIG. 2. Anteroposterior (left) and lateral (right) radiographs of the lumbar spine showing vertebral wedging in a 14-year-old boy with sickle cell anemia.

characteristic biconcave deformity called "codfish" vertebrae or "H-shaped" vertebrae. This typical appearance is a direct result of repeated microinfarctions in the central subchondral vertebral bone during childhood with resultant osteoporosis and intrusion of the nucleus pulposus of the disc. Vertebral collapse or wedging, or both, may also be present, secondary to osteoporosis. The degree of spinal involvement has been reported as being related to the duration and the severity of the disease.[4,8,10]

Crises are usually well managed with rehydration at 1.5 times the maintenance level, oxygen, analgesia, transfusion and recently hydroxyurea. This compound has been shown to increase the body's production of fetal hemoglobin, which does not contain a B-chain. It would therefore interfere with the crystallization process of sickle hemoglobin, thus reducing the symptoms of sickling.[12] The results of using hydroxyurea in 2 of our patients seemed to be promising.

Conclusions

Our study has emphasized the fact that sickling crises affecting the spine occur frequently in the pediatric age group with sickle cell disease. Back pain in children at risk for sickle cell disease should be considered as a possible sickle cell crisis and the child fully assessed for the presence of sickle cell anemia. Treatment is symptomatic for the back pain, which usually resolves with treatment of the sickle cell disease. Early results with the use of hydroxyurea to abort sickle cell crises appear promising.

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