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VERTEBRAL BONE COLLAPSE IN SICKLE CELL DISEASE: A REPORT OF TWO CASES
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VERTEBRAL BONE COLLAPSE IN SICKLE CELL DISEASE: A REPORT OF TWO CASES

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SUMMARY

We describe two female children both nine years of age with sickle cell anaemia and compression deformity of three successive lumbar vertebrae in one child and collapse of one lumbar vertebra in the other. Management for the two children included analgesics, antibiotics and application of a lumbar jacket for stabilisation of the spine. In both patients the vertebral bodies remodelling with re-generation of the tissue. This report is being made to highlight the improved chances of response with early detection and adequate management.

INTRODUCTION

Sickle cell haemoglobinopathy is common in Nigeria, affecting 2-3% of children(1,2). The disease process affects all the bones in the body and its effect on the vertebral bones have been described. Radiological features of these vertebral bodies include a coarse trabecular pattern, persistent anterior vertebral notching, biconcavity of the bodies, step ladder effect, massive collapse of the 'centra', and compression deformities(3,4). Most previous reports have focused on the pathological processes, none has looked into the management with a view of maximizing the reparative process and so prevent further damage(5-7). These case reports are presented to illustrate an aspect of management that has not been previously described for children with sickle cell anaemia.

CASE REPORTS

Case 1: A nine year old female, N C, presented with a one week history of waist pain, intermittent fever and inability to bend forward at the waist. The child's genotype was unknown. There was no history of trauma. The pain was dull in character and non-radiating. Fever which was worse in the evenings responded to antipyretics. Mobility at the waist was impaired and the child walked stiffly. There was no prior history of similar illness, bone pains or jaundice. She was fully immunised and had no history of contact with any one with tuberculosis.

On physical examination, at presentation, she was found to be of an asthenic build, mildly pale, anicteric, a febrile and not in any obvious distress. She was walking with a marked lumbar lordosis and a stiff gait. The muscle tone and power in the lower limbs were normal, the deep tendon reflexes were not exaggerated and sensation was intact. There was mild tenderness over the lumbar spine but no gibbus was discernible.

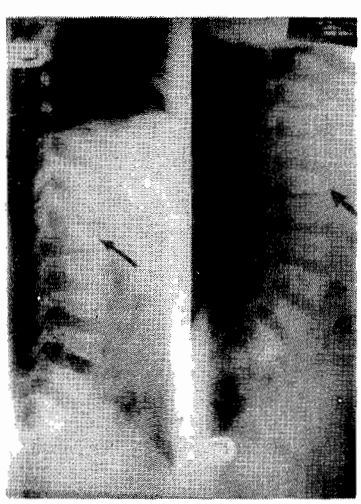
She had a splenomegaly extending 4 cm below the left costal margin. All other systems were essentially normal.

Laboratory investigations revealed PCV of 24% genotype SS, White cell count 10,400 mm^3 with a neutrophil of 68%, lymphocyte of 29% and oeosinophil of 3%.

The erythrocyte sedimentation rate was 35 mm in the first hour. There was no malaria parasite and mantoux result was 3 mm. Spinal radiograph (Figure 1) showed complete resorption of the anterior vertebral end plate of the residual box of L_1. There were areas of calcification within the dead bone with loss of L_{1-2} disc space and involvement of the contigous surface of the body of L_2. There was generalised thinning of the cortices of T_{12}-L_3 vertebral bodies with widened intervertebral spaces and thickening of the trabeculae with a resultant coarse bone texture. Biconcave discs were noted to varying extent throughout the thoraco-lumbar spine.

Figure 1

Lateral spinal radiographs showing vertebral body collapse of L_1 on the LEFT and remodelling of L_1 with loss of height on the RIGHT



She was managed with antimalarials, analgesics and a lumbar jacket was applied initially for six months. Removal resulted in a relapse needing a further two months of the jacket. Spinal radiograph done after 10 months showed L_1 vertebral body remodelling neatly with persisting L_{1-2} disc space loss and loss of height of the anterior portion of L_1 (Figure 1).

Twelve months after the lumbar jacket was removed she had no further problems.

Case 2: A nine year old female, 00 presented in this hospital with a three week history of waist pain with fever and a four day

had been made at the age of four years. The waist pain interfered with the child's mobility and was worsened by movement at the hips, lower limbs and by standing, but was relieved by the child lying down. The fever was high grade, intermittent and worse in the evenings. She was initially treated at the emergency room with intravenous fluids, analgesics, antimalarials and transfusion of one unit of packed cells. She was discharged after 96 hours but four days later the child was unable to sit up in bed or stand with or without support on account of the pain.

On admission into the ward she was found to be fairly well nourished with the typical sickle cell facies of bossing, gnathopathy and asthenia. She was pale, febrile with a temperature of 38.6°C and anicteric. She was fully conscious and alert, Kernig's sign was negative and the cranial nerves were intact. She had a gibbus over the lower spine and her lower limbs were hyptertonic with hyperreflexia, power was grade 4 and her sensation was intact.

There was a hepatomegaly of 8 cm below the costal margin, the liver was soft, non tender with a sharp edge and smooth surface.

Investigations disclosed anaemia with PCV of 23% Erythrocyte sedimentation rate of 150 mm in the first hour, white blood cell count of 17,500 mm^3 with neutrophil, 47% lymphocyte 51%, eosinophils 2%. Mantoux test was negative. Spinal radiographs showed multiple osteolytic lesions involving the bodies of L_{3-5} with loss of height of affected vertebral bodies. There was reduction in the disc space between L_{2-3} and widening of L_{3-4} disc space. The pedicles were intact. There was normal alignment of the vertebral body which also showed coarse traberculae. Blood culture grew *staphylococcus aureus.*

A lumbar jacket was applied and the child treated with intravenous antibiotics, initially, followed with oral antibiotics, for a total of six weeks, bed rest and analgesics. The lumbar jacket was removed after three months when the spinal radiographs showed marked healing of the lumbar vertebral bodies with marked regeneration of the bony tissue.

CONCLUSION

Most case reports on vertebral bone destruction in sickle cell haemoglobinopathy has focussed on the pathology and causative factors(4-7). There has been no report made regarding management of this condition. In view of the reported vanishing of the vertebral bone in a young male from this environment, possibly as a result of late presentation which prevented adequate management(6), we report these cases where management included application of lumbar jackets to stabilise their spines.

The ability of vertebral and other bones, in cases of sickle cell haemoglobinopathy, to regenerate has been noted by several authors(3,4). Application of a lumbar jacket might not necessarily hasten this process but would hasten ambulation of a patient and allow for outpatient management, especially in this environment where economic factors contribute greatly to patient care. It would also help in immobilising the spine of the patient and prevent further deterioration and loss of bone, thereby improving the affected patient's well being.

One wonders whether the patient, in the report by Ozoh et al(6) would have saved his vertebra had he been treated with a lumbar jacket. The discrepancy in the duration of application of jackets in these two patients could possibly arise from the causative factors (infection vs infarction) involved in the collapse of the vertebral bodies.

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