‘STEP VERTEBRAE’, ‘FISH MOUTH VERTEBRAE’ OR ‘H VERTEBRAE’ IN SICKLE CELL DISEASE

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ABSTRACT

Sickle cell disease is associated with damage to bony tissues, particularly in adults (1-3). The severity of the sickling condition, as well as genetic determinants such as that for annexin and steroid hormones such as vitamin D are being investigated epidemiologically to ascertain their relative significance. Most bony changes are not visible on clinical examination. We present an image from a teenager who presented with back pain and radiographic changes in his vertebrae.

KEYWORDS

Vertebra, Sickle cell, Fish-mouth, Step vertebra, H-shaped vertebra, bone Infarction

INTRODUCTION

A 16 year old boy with known sickle cell disease presented with intermittent pain in the lower back that had been troubling him for several weeks. Clinically he appeared anemic, icteric and distressed because of lumbar pain. There was no fever. His gait was slow but symmetrical. A clinical examination demonstrated muscle spasm and bilateral tenderness in the lower back. A careful neurological examination of the patient, particularly with respect to his legs, was normal. The plain radiograph collected showed a loss of lumbar lordosis. There are biconcave vertebral bodies. Critically, there is no narrowing of inter-vertebral disc spaces. A frequent location of vascular occlusion in sickle cell disease is the microcirculation located inside long bones, vertebrae, and ribs resulting in painful ischemic necrosis. It is postulated that following repeated episodes of ischemia and micro infarctions, the original concavity of the superior and inferior surfaces of the vertebral bodies becomes exaggerated, leading to this pathognomonic vertebra sign.

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This vertebral deformity has been referred to as the “fish mouth deformity” or “fish-vertebra sign”; some use the term ‘H-shaped’ vertebra (4,5). However these may be inaccurate as they suggest a pathophysiology similar to the collapse of weakened bone seen in osteoporosis. The term ‘Step vertebra’ is felt to be a more appropriate description based on the radiological appearances. The outer vertebral regions receive arterial blood from many branches of shallow perforating arteries while the central region is reliant upon branches of a single artery that courses through the marrow. Because of the anatomy of vertebral bodies, cumulative impedance of blood flow by sickled cells is more likely to have a greater effect on the central zone of the vertebral body. Step vertebra therefore develop following chronic microvascular infarctions by sickled red blood cells that occlude the single arterial supply to the central area of the vertebral bodies, resulting in defective bone development (6). In the short term this may require analgesia, in the long term mechanical or orthopaedic approaches may be indicated.

The patient recovered from his back pain over several weeks. Checks of his vitamin D showed this to be mildly insufficient; his calcium levels, alkaline phosphatase and parathyroid hormone level were normal, as was his
zinc level. He had no signs of iron overload and his ferritin was in the normal range (7,8). However from the perspective of his sickle cell disease he had suffered a haemolytic picture with consistently raised LDH and bilirubin, a raised reticulocyte count and low haemoglobin for several years. As a younger child his condition had been characterized by regular crises, but normal transcranial Doppler measurements and no evidence of any end organ damage. The patient and his family had been unhappy to commence the use of hydroxyurea for several years prior to his presentation with back pain. The degree of bone fragility or reduction in bone density in patients such as this merits formal assessment: this is planned.

CONCLUSIONS

Vertebral changes are often stated to be rare in adolescent patients with sickle cell anaemia. In this particular case it may represent a manifestation of severity of the underlying haemoglobinopathy, given that other checks of his bone health appeared unremarkable. His presentation challenges clinicians: should we be screening for spinal changes earlier in adolescent patients with sickling conditions?

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REFERENCES