

Osteopenia in Crohn's disease

Fiona J Cowan, David R Parker, Huw R Jenkins

Abstract

A 16 year old boy presented soon after diagnosis of Crohn's disease with diffuse osteopenia of the lumbar spine with collapse of three vertebral bodies and grossly reduced bone mineral density noted on dual energy x ray absorptiometry scan. Treatment of the underlying Crohn's disease and induction of puberty resulted in marked improvement in bone mineralisation.

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Keywords: osteopenia, bone mineral density, Crohn's disease, pubertal delay.

Osteopenia with severe clinical consequences has been widely reported in adults with inflammatory bowel disease.¹ However, very little has been reported of bone mineralisation in children with the disease. One study has shown that older children with Crohn's disease are at risk of diminished bone mineralisation which correlated with depressed body mass index, increased disease activity, and high dose steroid treatment.² This case report describes a child with severe osteopenia and vertebral collapse noted soon after diagnosis of Crohn's disease.

Case report

A boy aged 16 years was referred by his general practitioner with a four year history of anorexia, poor weight gain, and poor growth. His stool frequency was five times/day but he had passed no blood or mucus in the stools. He had lost 4 kg over the six months before his referral and he weighed 24.15 kg. His height was 140.1 cm (4.7 SD below the mean) and his height velocity was 0.5 cm/year. On examination he was clinically anaemic and cachectic and he had finger clubbing. He was prepubertal (pubic hair stage 1, genitalia stage 1, testes 4 ml bilaterally). Examination was otherwise unremarkable. His bone age was delayed at 11.4 years and gross osteopenia was noted on radiography. Results of investigations were; haemoglobin concentration 74 g/l, platelet count $741 \times 10^9/l$, albumin concentration 19 g/l, erythrocyte sedimentation rate 5 mm/hour.

A barium meal revealed two narrowed, ulcerated segments in the proximal jejunum and the terminal ileum. A diagnosis of Crohn's disease was made and prednisolone treatment was commenced at 2 mg/kg/day and a percutaneous gastrostomy tube inserted for elemental feeding (Elemental 028, Scientific Hospital Supplies). The steroid dose was quickly reduced as he responded well to the elemental diet and one month after diagnosis he was on

an alternate day regimen of 0.5 mg/kg. Colonoscopy was macroscopically and histologically normal. Three months after his initial referral he began to complain of severe back pain and became unable to weight bear.

Radiographic examination of his spine revealed diffuse osteopenia of all bony structures with reduced height of the lumbar vertebrae and wedge fractures at T12, L1, and L2. A whole body dual energy x ray absorptiometry (DEXA) scan was performed on the Hologic QDR 1000/W scanner and revealed a total bone mineral density (BMD) of 0.8 g/cm² (3.8 SD below the mean for normal young males) and the lumbar spine BMD was 0.3 g/cm² (-6.5 SD). Even allowing for his weight and pubertal delay by comparing the lumbar spine BMD with that of a child with similar bone age it is 4.8 SD below the mean. A luteinising hormone/follicular stimulating hormone stimulation test showed a prepubertal response. Prednisolone treatment was reduced and stopped over the next three weeks. He was commenced on testosterone undecanoate 40 mg on alternative days and continued the elemental diet. Over the next 11 months his growth velocity improved to 9.9 cm/year and he progressed through puberty, achieving Tanner stage III. Repeat DEXA scan 11 months after diagnosis of osteopenia showed great improvement with a significant increase in bone mass and lumbar spine BMD had increased to 0.46 g/cm², an increase of 74.5%, although the total BMD remained low at 0.743 g/cm². Fourteen months after diagnosis he is asymptomatic, his haemoglobin, platelet count and albumin are normal, his weight has improved to 42.4 kg and he has achieved a height of 149.5 cm (-3.7 SD).

Discussion

Growth retardation and pubertal delay are well recognised complications of inflammatory bowel disease in childhood. The reported frequency of growth impairment in these children varies from 32 to 88%.³ In addition, an increased rate of cortical bone loss has been found in adults with inflammatory bowel disease.¹ Several inter-relating factors may have given rise to the osteopenia and growth retardation observed in our child. Malabsorption, protein energy deprivation secondary to a poor diet, and inadequate calcium intake will all result in a reduced rate of bone production.⁴ Furthermore, steroid treatment has a direct inhibitory effect on bone production and also results in an increased rate of bone loss by inducing hypercalciuria. In view of the early finding of osteopenia it may have been inappropriate to instigate high dose steroid treatment as

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elemental diet alone has been shown to be very effective in inducing remission in childhood Crohn's disease.⁵ In our child, however, the contribution of steroids to the bone disorder was likely to be minimal in that osteopenia was present in bone age radiographs and DEXA scans early in the illness after a very short duration of treatment. Sex steroids are known to be important mediators of ossification and of the pubertal growth spurt. Our child had evidence of both malnutrition and delayed puberty both of which are likely to have contributed to the poor bone mineralisation as BMD is known to increase with weight and pubertal Tanner stage and these have been found to be the best predictive indicators of bone mass and BMD.⁶ The marked improvement in our child's lumbar spine BMD was achieved by both improving his nutritional status by controlling the underlying Crohn's disease and by inducing puberty.

The lesson to be learned from this case is that gross osteopenia may be present at the time of diagnosis and it is therefore important to exclude it in all children with Crohn's disease. We suggest that the bone mineralisation status of all our children with inflammatory bowel disease should be determined at presentation and at regular intervals throughout their treatment.

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Dopa responsive dystonia

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Abstract

There may be insufficient awareness of dopa responsive dystonia (DRD), which has a characteristic diurnal variation of symptoms. Two children are reported in whom the diagnosis of DRD was missed. The first was thought to have hysteria and the second hereditary spastic paraparesis. A full history is vital for the diagnosis of this important treatable syndrome.

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Keywords: dopa responsive dystonia.

Segawa *et al* in Japan reported a group of children with progressive idiopathic dystonia where the symptoms deteriorated through the day and improved after sleep.¹ There was complete resolution with a small dose of levodopa. A gene for this disorder, inherited as a dominant, has been recently mapped to 14q.²

The differential diagnosis includes other dystonias, juvenile onset parkinsonism, spastic and psychogenic disorders.^{3 4} We describe two patients to illustrate early recognition and differential diagnosis of dopa responsive dystonia (DRD). We particularly emphasise the need to consider the diagnosis in a child presenting with a progressive, apparently hysterical or spastic, gait disorder.

Case reports

PATIENT 1

This 6 year old boy was referred by an

orthopaedic surgeon. He had presented at 5 years old with a three month history of walking on his toes, particularly on the left side. His symptoms had been worsened by a plaster of Paris splint. After referral to the paediatric department the bizarre nature of his gait initially suggested a hysterical disorder.

His symptoms had worsened coincident with physical activity during the summer holidays. His difficulty walking was asymmetrical, with stiffness of the left leg. Running was easier than walking. We suggested the diagnosis of DRD when his parents reported that he had no symptoms in the morning, could walk to school without difficulty, but required physical support to walk home, and in the evening could not walk more than a few paces unaided.

On examination he walked, slightly stooped forward, with a stiff gait on his toes on the left with some stiffness of his left knee. He had associated movements of both upper limbs when walking, with abduction of the arms, flexion of the elbows, pronation and flexion of the wrists and metacarpophalangeal joints, and extension of the fingers. Lower limb tone was mildly increased, more marked on the left, with brisk reflexes, ill sustained bilateral ankle clonus, and normal plantar responses. Despite his difficulties with walking he could run, jump, and hop. Radiography of his hips, computed tomography of the head, and copper studies were all normal and there were no Kayser-Fleischer rings.

He responded excellently to a small dose of levodopa (25 mg) and carbidopa (6.25 mg) (Sinemet LS). Within a week, he had

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