“Tower vertebra”: a new observation in sickle cell disease

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Abstract Background. Skeletal abnormalities are common in sickle cell anemia. Ischemia, infarction, and growth disturbance of the thoracic and lumbar vertebral bodies are among the most common abnormalities, and can suggest the diagnosis radiographically. Design and patients. We recently encountered two adult patients in whom vertebrae had grown abnormally in height adjacent to infarcted short vertebrae. We then reviewed the thoracic and lumbar spine radiographs of 54 more adult patients with sickle cell anemia. Results and conclusion. A total of eight patients (14%) displayed infarcted vertebrae with compensatory vertical growth of at least one adjacent vertebrae. These resemble the elongated vertebral bodies associated with other conditions. We can find no prior report of this finding in association with sickle cell anemia. Key words Sickle cell anemia · Vertebral body · Vertical growth

Introduction

The most frequent clinical finding in sickle cell disease is bone pain. Bone infarction secondary to microvascular occlusion is frequent, and vertebral infarction is one of the commonest sites. Two patterns of vertebral ischemia collectively called “Reynold’s vertebrae”, are considered diagnostic for microcystic anemias. These are the so-called fish-vertebra and the H-vertebra. We describe here the compensatory increase in vertical height of thoracic vertebrae in response to childhood infarction, and subsequent hypoplasia of adjacent vertebrae. To our knowledge this is a previously unreported finding. We suggest that sickle cell anemia be added to the possible pathologic mechanisms for elongated vertebral bodies. Also, “tower vertebra” should be added to the list of radiographic findings that should suggest sickle cell anemia.

Methods

After noticing the deformity in two patients initially, we reviewed the radiographic records of 67 additional patients seen in the Adult Sickle Cell Anemia Clinic at our institution. Thoracic and lumbar spinal radiographs were available for review in 54 of these. All patients were physiologic adults, greater than 17 years of age, with no evidence of continued epiphyseal growth. All patients had well-documented, chronic disease, many with severe courses. Eleven patients without thoracic or lumbar spine films were excluded. We reviewed the spine films for evidence of vertebral infarction: Reynold’s vertebrae (both “fish” and “H” varieties), and “towering”. “towering” was identified by measurement of vertebral heights, comparison with adjacent vertebrae above and below, and comparison with means established by Hurxthal [1]. “Tower vertebrae” were further
characterized as having concentric hypertrophy, hemihypertrophy, or central “ballooning”. Clinical records were reviewed.

Results

Of the 56 patients with thoracic or lumbar spinal radiographs, 52 (92%) had radiographic evidence of spinal ischemia. Thirty-eight (67%) displayed Reynold’s vertebrae, 17 (44%) showed fish vertebrae, and 31 (81%) showed H vertebrae. Ten (26%) showed evidence of both deformities on the same film. Eight (14%) of these patients showed evidence of the tower vertebra deformity, with two (3%) showing concentric hypertrophy, three (5%) showing hemihypertrophy, and three (5%) showing central ballooning. Two (3%) patients displayed towering of more than one vertebra in the spinal column. One patient showed confluent towering of L1 through L4 with adjacent infarction of the entire thoracic vertebral column.

In our series, tower vertebrae were found exclusively next to adjacent infarcted vertebrae. We found through careful measurements that the tower vertebrae are taller than normal vertebral bodies based on mean heights established by Hurxthal [1]. These measurements also revealed that the adjacent infarcted vertebrae were shorter than average. This combination makes tower vertebrae appear exceptionally tall. All the patients with positive findings were female and homozygous for the sickle cell trait.

Discussion

Sickle cell anemia is a genetic disease of red cells that results in crystallization of malformed hemoglobin S in low oxygen states, secondary marked distortion of red cell architecture, resultant occlusion of the microvasculature, and infarction of underperfused tissues. Red cells are hemolyzed due to sickling leading to chronic anemia.

Bone involvement is present in about 80% of patients with sickle cell anemia, and produces the majority of symptoms in the disease. Two forms of pathology produce these symptoms. Thrombotic microvascular infarction of bone can lead to acute and chronic bone pain, crisis, and occasionally osteomyelitis. Chronic anemia elevates erythropoietin levels, and leads to expansion of the marrow cavity, resulting in marrow hyperplasia [2].

Necrosis of the femoral head and secondary osteoarthritic changes are the most common bone findings on radiographs. However, some spinal changes on radiographs are highly suspicious for sickle cell anemia and can suggest the diagnosis. These include demineralization, generalized infarction with increased density, and changes in vertebral bodies due to ischemia. The pathognomonic ischemic vertebral findings first described by Reynolds are divided into two categories: the fish vertebra is a smooth biconcave deformity secondary to marrow hyperplasia with resultant osteoporosis, while the H vertebra is an abrupt cup-like lesion due to central infarction and collapse [3, 4]. More unusual findings reported include anterior vertebral vascular notches, [5] and the “vanishing vertebra” [6]. The tower vertebra has not been reported in association with sickle cell anemia.

Tower vertebra is an increase in vertical height of a vertebral body without an appreciable increase in girth. It is associated with several conditions, including diseases of connective tissue, vertebral fusion, and prolonged inactivity. Marfan’s syndrome is a connective tissue disorder of fibrillin. It is associated with elongation and hyperflexibility of the entire musculoskeletal system, including the vertebral column. Congenital block vertebra, Klippel-Feil syndrome, developmental fusion (i.e., Scheuermann’s disease), and tuberculous spondylitis result in tower vertebra. This is due to ossification across the disk space, with no true lengthening of an individual vertebral body. Neuromuscular disorders (i.e., polio, myasthenia gravis) and prolonged inactivities create a developmental environment in which the growing vertebrae are allowed to lengthen unchallenged by gravity. Infarction of an adjacent vertebrae is a previously unreported cause of compensatory heightening of vertebrae.

We offer two possible explanations for this new finding. The first involves compensatory enlargement of body tissues. Several organ systems, including liver, kidney, and bone, have been shown to increase in size or length in response to trauma, removal, or disease. Regeneration of human liver occurs after cirrhosis, hepatitis of all types, and surgical resection following trauma [7]. Enlargement of the contralateral kidney after nephrectomy, or other loss of renal tissue, is due to both hypertrophy and hyperplasia. A single kidney can assume 90% of the mass of both kidneys within 40 days [8]. Compensatory lengthening of bone in response to injury has also been shown to occur in children. After femoral fracture, children display overgrowth of the broken femur which averages 0.92 cm. Additionally, 82% of patients develop overgrowth of the ipsilateral tibia averaging 0.29 cm [9]. It seems that the body recognizes normal parameters for metabolic and structural activity, and contains intrinsic corrective mechanisms to compensate for loss or injury. We believe the tower vertebra may be an attempt to maintain pre-set height expectations mechanically, similar to bone lengthening after femoral fracture. The loss in height of an adjacent vertebrae may trigger vertical hypertrophy of adjacent levels.

The second possible explanation for the finding of tower vertebra involves marrow hyperplasia. The body responds to chronic anemia by increasing erythropoietic tissue mass. One manifestation of this pathology is marrow hyperplasia, an increase in the size and metabolic rate of the marrow cavity. Marrow hyperplasia has been documented in 42% of children with severe sickle cell