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CLINICAL IMAGE

Bone within bone in juvenile dermatomyositis

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Presentation

Twenty-eight-year-old woman with juvenile dermatomyositis (JDM) since 7 years old who was taking immunosuppressive therapy (azathioprine 100 mg/day and mycophenolate mofetil 3 g/day) presented as an asymptomatic patient for a rheumatology consultation. A previously ordered lumbar spine X-ray (Fig. 1) shows a "bone within bone" appearance.

The patient did not previously use bisphosphonates. She had normal psychomotor development and she had no history of neoplasms. Additional investigation was carried out, and the patient also showed no changes in blood count, kidney or liver function, alkaline phosphatase, serum calcium, phosphorus/vitamin D, or bone densitometry.

Discussion

The "bone within bone" aspect is characterized as a rare radiographic appearance where one bone appears to arise within another bone [1]. This aspect is commonly described in childhood. It can occur due to failure to inhibit osteoclast activity, crystal deposition, pathological periosteal bone formation, or subcortical osteopenia [2, 3].

Previous studies have shown osteometabolic alterations in rheumatic patients that may be the key to this condition: (i) decrease in serum levels of osteocalcin; (ii) the chronic inflammatory process in JDM with increased interleukin (IL)-1, IL-6, IL-7, IL-17, and tumor necrosis factor alpha (TNF-a); (iii) increased serum levels of receptor activator of nuclear factor kappa-B ligand (RANKL) with decreased levels of OPG (osteoprotegerin); and (iv) chronic use of glucocorticoids and low levels of serum calcium and vitamin D. Even with a decrease in the response to the absorption of serum calcium in the diet [4]. These alterations associated with genetic factors such as sex (female gender) and environment–low socioeconomic status (due to lack of access to adequate food and medical assistance), and late onset of puberty present in patients are described as possible triggers for worsening bone health in patients with idiopathic inflammatory myopathies (IIMs) [1, 4].

Furthermore, it is important to point out that the increase in IL-6 and TNF-a in patients with rheumatic diseases in childhood may lead to changes in bone growth because (i) it decreases IGF-1 (insulin-like growth factor 1) in animal models, (ii) the increase in C-reactive protein (CRP) has already shown an inverse correlation with bone mineral density, and (iii) decreased muscle mass, probably associated with late puberty due to chronic disease [5]. Besides that, the rule in JDM is not clear.

In the case of our patient, we believe that due to the chronic inflammatory process and the use of glucocorticoids at a very young age and in high doses can justify the changes in bone growth.

Differential diagnoses that should be considered with the radiographic appearance displayed include the following: (i) physiological changes in bone formation in children; (ii) use of medications such as bisphosphonates or prostaglandins E1; (iii) genetic disorders such as osteopetrosis, hereditary hyperphosphatasia, Caffey's disease, Paget's disease, sickle cell anemia, and Gaucher disease; (iv) neoplasms such as leukemia and metastases or postchemotherapy; and (v) iatrogenic or spurious causes [6]. Radiological differential diagnosis can be made with "picture frame vertebral body" and "sandwich vertebral body" present in Paget's disease and "rugger jersey spine" in hyperparathyroidism [2, 3]. Treatment depends on the underlying cause.

Thus, this is the first description of "bone within bone" image in a patient with juvenile dermatomyositis and idiopathic inflammatory myopathies in general. It seems, despite being rare, JDM can occur with this aspect.

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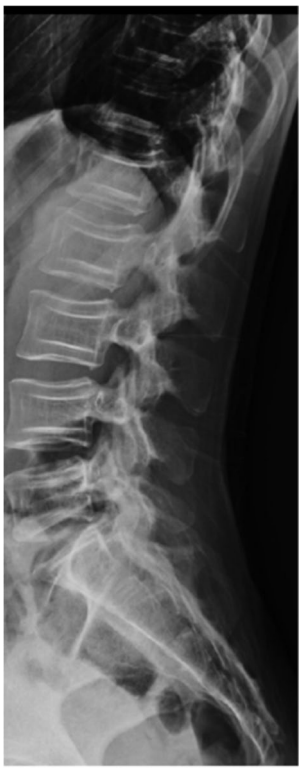

Fig. 1 Radiological appearance of Bone within Bone in the lumbar spine

Author contribution All authors contributed equally to this paper.

Declarations

Disclosures None.

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