CASE REPORT

Pain and osteolysis of the thoracic spine - A case of a rare monostotic fibrous dysplasia manifestation

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Abstract

We describe a case of a young farmer from Central Macedonia, Greece suffering of a mild back pain more than one year. His medical history included hypercholesterolaemia (IIa type) and two episodes of spontaneous pneumothorax of unknown origin two and three years ago respectively. A full imaging survey revealed a single osteolytic lesion at the seventh thoracic vertebra. A CT guided needle biopsy was performed. Diagnosis based on clinical, imaging and histological findings was monostotic fibrous dysplasia of the thoracic spine. We discuss the clinical features and treatment of this non neoplastic condition which may simulate bone osteolytic tumor. Furthermore a possible correlation of concomitant conditions existing in our patient such as the metabolic disorder of hypercholesterolaemia and especially the history of spontaneous pneumothorax episodes with fibrous dysplasia within the spectrum of connective tissue disorder is discussed. Hippokratia 2008; 12 (4): 254-256

Keywords: monostotic fibrous dysplasia, osteolysis, thoracic spine, spontaneous pneunothorax

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Case Report

A 29-year old Caucasian man, farmer from Northern Greece was admitted in AHEPA Hospital because of a mild low and middle back pain lasting more than one year. No history of injury was reported. Generalized fatigue was referred while back pain was worsened during and after working in the farm. The symptoms were relieved after rest and after non-steroid anti-inflammatory agents (NSAID) administration. No morning or after rest stiffness was noticed. On clinical examination, the patient experienced tenderness on the mid -dorsal spine region, spreading to both sides from the lower cervical to lower thoracic level. No skeletal malformations, skin manifestations or neurological signs were noticed. There was a history of two episodes of spontaneous pneumothorax of unknown origin two and three years ago each and of known hyperlipidaemia (IIa type).

Laboratory tests revealed hypercholesterolaemia (cholesterol 350 mg/dl, triglycerides 135 mg/dl, LDL: 290 mg/dl). Inflammation markers (erythrocyte sedimentation rate, C-reactive protein), markers of bone metabolism (Ca, P, alkaline phosphatase, parathyroid hormone), cancer markers (AFP, PSA, CA10, CEA), thyroid hormones levels and immunological examinations (C3, C4, ANA, AMA, ASMA, anti-DNA, p-ANCA, c-ANCA, anti-RNP, protein immunoelectrophoresis) were normal.

A full imaging survey was performed including x-rays of the chest (F + P), the skull (F + P + Town view), the thoracic spine, the pelvis and the long bones without abnormal findings. Computed Tomography (CT) of thoracic spine showed a large lytic lesion of the left transverse process of the seventh thoracic vertebra (Figure 1A, 1B). MR Imaging (MRI) confirmed the CT findings with low signal in T_1WI and high signal in T_2WI. The bone scanning was not conclusive.

A tissue sample was taken from the T 7 vertebra with a CT-guided needle biopsy in order to clarify the nature of the osteolytic lesion. The specimen was embedded in paraffin blocks and stained with hematoxylin-eosin. The histological examination demonstrated spicules of new bone formation with interventing cellular fibrous tissue and calcifications. Immature bone showed no concentricity and was unable to get organized in normal bone (Figure 2). A zone consisted of thin connective tissue surrounded the whole lesion. Atypical cells and in general indications of malignant change were not found. According to clinical, imaging and histological findings a differential diagnosis between non - ossifying fibroma, neurofibromatosis, fibrous dysplasia (FD) and fibrosarcoma was performed whereas the final diagnosis was monostotic FD with a single manifestation at the thoracic spine.

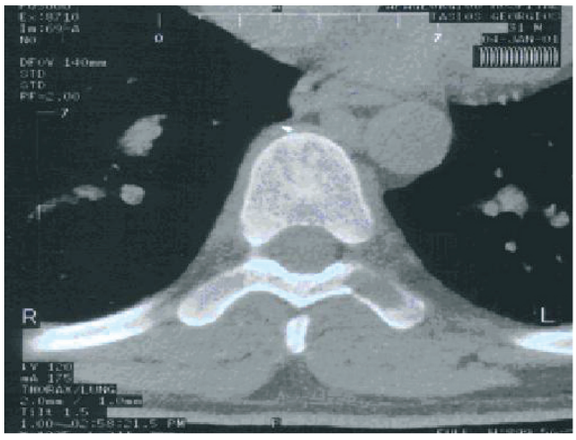

Figure 1A: CT of thoracic spine: A lytic lesion on the transverse process of the 7th thoracic vertebra.

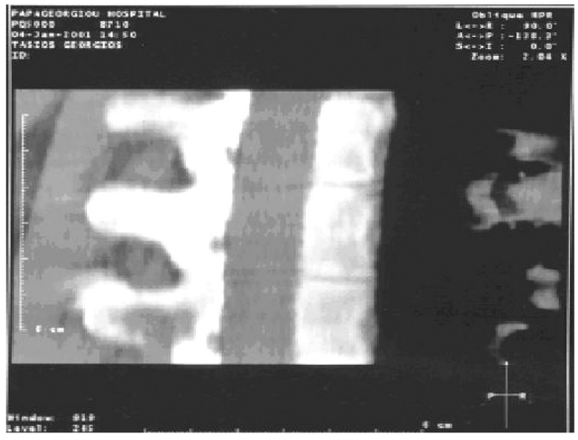

Figure 1B: CT of the thoracic spine (sagittal view) showing lytic lesion of the left transverse process of the 7th thoracic vertebra.

Discussion

FD is a benign disorder of bone in which proliferating fibrous tissue replaces the bony spongiosa. It is a clinical entity, which affects both sexes with equal frequency and presents at any age, the mean being 32 years, although it is usually diagnosed in childhood[1]. Depending on the extent of the disease (single or multiple skeletal sites), the manifestation can vary greatly. Monostotic FD is rare and appears at 20 - 30 year of age while polyostotic type is usually appears at 3 - 15 year of age. Monostotic FD of the thoracic spine is exceedingly rare[2-4].

It has to be emphasized that diagnosis of this non neoplastic condition which may simulate bone osteolytic tumor, can often take place incidentally, because small osteolytic lesions may be soundless for a long time[4]. Our notifications are in accordance with recent literature[1-5].

It is worthwhile to notice that FD of the bones can clinically be manifested as part of the McCune-Albright syndrome. Endocrine dysfunction skin lesions and FD are the sound clinical points of the syndrome. Endocrine dysfunction may be seen as precocious puberty[6,7]. A hypophyseal adenoma secreting prolactin and adrenocorticotrophic hormone (ACTH) may also be included resulting in acromegaly, gigantism and/or hyperprolactinemia. Cutaneous "cafe-au-lait" spots are, on the other hand, are the most common skin lesion featuring this syndrome[8]. In our case, we did not notice any of these features, something concomitant to the fact that polyostotic and not monostotic FD is frequently correlated with skin lesions and endocrinopathies[1]. On the other hand, it was of great interest that our patient had suffered two incidents of spontaneous pneumothorax in the near past. We did not find any correlation in the literature between FD and spontaneous pneumothorax but a possible explanation based on a diffuse disturbance of the connective tissue is attractive. Furthermore the presence of high plasma levels of cholesterol, which is a major component of the cell membrane, implies a metabolic disorder. The latter has to be investigated for possible correlation with a connective tissue disorder such as FD. Both concomitant conditions could be related with FD within the spectrum of connective tissue disorders. A case report of anorexia nervosa has also been described, in which malnutrition caused connective tissue changes leading to diffuse structural connective tissue derangement leading to spontaneous pneumomediastinum and diffuse soft tissue emphysema (subcutaneous, epidural and retroperitoneal)[9].

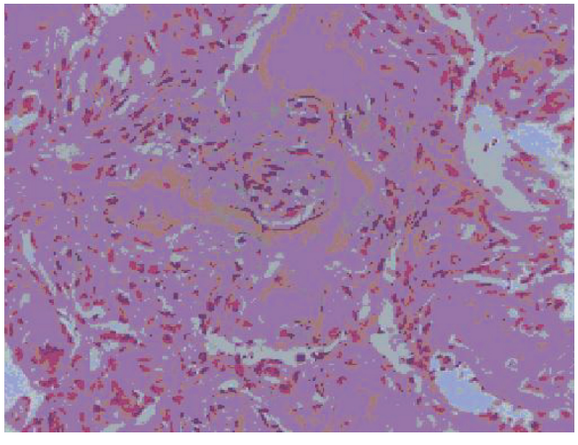

Figure 2: Immature abundant trabecular bone formation with bland-looking fibrous tissue surrounding it. The fibrous stroma consisted of spindly to oval fibroblasts scattered within eosinophilic collagen (HE X160).

It is well recognized, that FD is caused by activating an autosomal mutation of the Gs alpha subunit of protein G resulting in an increased c-AMP concentration and thus in retarding of osteoblast differentiation with abnormal bone formation. There is also an increase in interleukin-6 levels expressing bone resorption, which is the rationale for administering bisphosphonates in the management of these patients[1,6,7]. Some complications, such as nerve compression or malignant change in FD are uncommon. FD may undergo malignant change of osteosarcoma,

chondrosarcoma, fibrosarcoma or mimics them. A review in the literature reveals 101 cases of FD complicated by malignant change[6,10]. The differential diagnosis is based on plain X- rays, CT scan or MRI, but mainly is established by the histological confirmation. Although it is rare, the exclusion or the presence of malignant change remains the vast diagnostic criterion. Therapy aims in removing the lesion only where necessary (malformation, pathological fractures, nerve compression or deformity). Bromocryptine and bisphosphonate pamidronate failed in producing positive results due to relapse or adverse effects during treatment and leading in early suspension of treatment[11]. Actually, as pain is the common occurrence in FD[12], a step-wise approach to its management, starting with NSAID is recommended in symptomatic patients. Although NSAID are used mainly, at times that pain can be severe, use of narcotic analgestics is required for adequate control, also according to the guidelines of the Agency for Health Care Policy and Research, the Joint Commision on Accreditation of Health Care Organizations and the American Medical Society. On the other hand, pain prevalence in patients with FD, its association with skeletal sites and/or severity of the disease, as well as its treatment are poorly understood yet[1,13,14].

In our patient, there was no indication for surgical intervention, and treatment according to the above recommendations was conservative consisting on administration of anti-inflammatory agents on demand. Clinical and laboratory follow up was recommended just in case of malignant change. Our patient is free of "lytic" lesions according to annual control with plain radiographs for more than four years.

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