Multiple painful bone cysts in a young man

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Clinical history

A 32 year old white man complaining of pain in the sacrum, lumbar, and right inguinal regions was referred to our service with an initial diagnosis of ankylosing spondylitis. Relevant medical history included removal of a mass in the neck 10 years previously, but the pathological diagnosis was not known. There was no family history of illness.

Physical examination of the neck revealed a soft, mobile, painless tumour, 5 × 3 cm in size in the left side of the neck, just below the sternocleidomastoid muscle. This tumour had been growing progressively for the previous six months. Mobility of the spine and peripheral articulations showed no abnormalities. There was no osseous tenderness. Laboratory data were not remarkable.

signal characteristics were also noted at the L4 level. Coronal views of the right acetabulum showed high signal intensity cysts (fig 5).

Differential diagnosis

The interesting feature of this case is the presence of multiple, well defined osseous lytic lesions involving the axial skeleton. The table lists the wide range of causes of multiple osseous cysts,^{1–3} which we have divided into two groups according to their frequency of occurrence.

Among the 'common causes' of multiple lucent lesions are *multiple myeloma* and *osseous metastasis*. These two diseases produce involvement of the flat bones such as the pelvis with a well defined profile, but without reactional

Radiological findings

Plain film radiographs (fig 1) showed normal sacroiliac joints, and a degenerate right hip joint with a subchondral cyst lesion with a sclerotic border, on the acetabular roof. The vertebral bodies of L1, L2, and L4 displayed increased vertical striations ('corduroy' vertebra) (fig 2). Computed tomography of the pelvis (fig 3) showed multiple intraosseous, well circumscribed lytic areas involving the ilium, sacrum, L5 vertebra, pubic rami, and femoral neck. No matrix calcification was evident. Magnetic resonance imaging of the lumbosacral spine displayed multiple vertebral body involvement predominately, with low (T1) and high (T2) signal intensity (fig 4). Mixed

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Figure 1 The degenerative subchondral cyst lesion with sclerotic border on the right acetabular roof is caused by degenerative disease. There are multiple lucencies in the pubic rami, with some expansion.

Figure 2 Increased vertical striations in L1, L2, and L4 vertebral bodies.

Figure 3 Computed tomography of the pelvis, showing multiple bone lesions affecting ilium, sacrum, L5 vertebra, pubic rami, and femoral neck.

Figure 4 Low (T1, left) and high (T2, right) signal characteristics of lumbar vertebral lesions seen on magnetic resonance imaging.

sclerosis or radiodense matrix inside. Lymphomas commonly produce a mixed lytic and sclerotic appearance. ‘Brown tumours’ of primary and secondary hyperparathyroidism

Figure 5 High signal intensity of multiple iliac cysts’ on T2 weighted magnetic resonance imaging scan in the parasagittal plane.

produce multiple small cysts in the pelvis, ribs, and long bones, associated with more characteristic radiological images of osteopenia, with subchondral and subperiosteal resorption. Primary and secondary amyloidosis are rare but well known causes of multiple lucent bone.

The ‘less frequent causes’ include *histiocytosis X*, all of the variants of which (for example, eosinophilic granuloma) produce cysts surrounded by sclerosis in the pelvis, skull, vertebrae, and long bones. In *Gaucher’s disease*, the cysts accompany other radiological images such as ‘Erlenmeyer’s flask’ deformity of the femora, or aseptic necrosis. Osseous *fibrous dysplasia* is a congenital disease in which fibroblastic cellular proliferation produces large bone cysts with a ground glass appearance and characteristic deformities. Osseous *enchondromatosis* is characterised by cysts in the tubular bones of the hands and feet, and metaphysis of the long bones with intra-tumorous opacities reflecting calcification or ossification of the cartilaginous tissue. *Diffuse cystic angiomas* produces multiple lytic lesions of bone that may affect the viscera and soft tissues.

Diagnosis

Cervical computed tomography revealed a homogenous left sided solid tumour in the soft tissues, near to vascular elements (fig 6). This was removed surgically, and a well capsulated, multiloculate mass with a cystic appearance and a milky content was found. Histologically (fig 7), there were numerous empty spaces, lined with smooth endothelium, that stained positively with factor VIII associated antigen.

Differential diagnosis of multiple lytic bone lesions

Common causes
Multiple myeloma
Metastasis
Lymphoma
‘Brown’ tumours
Amyloidosis
Less frequent causes
Histiocytosis X
Gaucher’s disease
Fibrous dysplasia
Enchondromatosis
Diffuse cystic angiomas

Figure 6 Computed tomography of the neck, showing solid and homogeneous tumour on the left side.

Figure 7 Histological section showing dilated lymphatic channels of irregular size and shape, lined by endothelium.

Figure 8 Cavernous angioma of bone marrow: large cysts lined by endothelial cells replacing haematopoietic tissues.

Figure 9 Splenic hypodense images on abdominal computed tomography.

The anatomopathological diagnosis was lymphangioma.

Some months later, a bone biopsy specimen obtained from the right ilium (fig 8) revealed the existence of multiple osseous cysts, some with a lymphatic content, surrounded by a layer of flat endothelial cells that stained positively with factor VIII associated antigen. The final diagnosis was *diffuse cystic angiomatosis*. Computed tomography of the abdomen searching for visceral foci of disease displayed several hypodense images in the spleen, suggestive of angiomas (fig 9).

Discussion

Cystic angiomatosis is a rare pathological entity of vascular origin that is characterised by osseous lesions frequently combined with angiomatous involvement of the viscera and soft tissue.⁴⁻⁷ The aetiology is unknown; it could be a true neoplasm or, as the majority of authors believe, a congenital malformation: a vascular hamartoma.⁸⁻⁹ It is more common in men, with half of the cases appearing at puberty, and most of them revealed coincidentally during radiological examination. Up to 1984, the number of published cases of diffuse angiomatosis had not exceeded 100.⁷

Histologically, angiomatosis consists of multiple dilated vascular canals, of cystic appearance and with walls lined with a layer of flat endothelial cells. The cysts may contain blood or a proteinaceous fluid, which often makes it impossible to determine if the dilated canals are related to a haemangioma or a lymphangioma;¹⁰ patients may in reality have a lymphangiomatous lesion at one site, and a typical haemangioma at another. Because of this, the name ‘cystic angiomatosis’ is preferable.

In the soft tissues, the angiomas are usually located in the neck or axillary regions. Visceral involvement, usually of the lung, and less frequently the spleen, liver, or mediastinum, reaches 60–70% in all cases. The condition carries a variable prognosis; some cases produce death in infancy.¹¹⁻¹² Osseous lesions produce symptoms when a pathological fracture occurs but, as in the patient reported here, osseous pain may exist without fracture, mimicking arthropathy, and rheumatologists may be left to deal with the problem. When the condition is confined to the skeleton, the prognosis is good, as the lesions tend to remain static. Soft tissue and visceral involvement such

as occurred in this patient are associated with a more serious outcome.

Radiological changes^{13 14} appear in the pelvis, femora, skull, and ribs; the humerus, scapulas, clavicles, and spine are frequently also affected, but the hands and feet rarely so. Radiologically, the multiple osseous areas in the bone have well defined edges and are surrounded by a sclerotic ring with a honeycomb appearance. The intramedullary 'cysts' are round or oval, and the cortex may appear expanded without breaking unless a pathological fracture occurs. Periostitis is not frequent.

Angiomas are variable in appearance on magnetic resonance imaging. On T1 weighted images, the signal intensity varies from low to high, depending on the amount of adipose tissue present and the predominant tissue components—haemangioma or lymphangioma. T2 weighted images usually show areas of very high intensity corresponding to the vascular or fluid components. In the patient we describe, and in another recent report,¹⁵ the magnetic resonance signal characteristics differed somewhat from those of vertebral haemangiomas, with which high signal intensity is seen on both T1 and T2 weighted sequences.¹⁶

Summary

To confirm the diagnosis of diffuse cystic angiomas, it is necessary to biopsy the bone: the walls of the cysts react to immunological markers of the endothelium (antigens related to factor VIII and CD31)¹⁰. Alternatively, lymphography can avoid the necessity for biopsy.¹⁷ Evolution of the condition is variable and depends on the extent of visceral

involvement, and usually the extent of soft tissue and visceral involvement dictates the morbidity and mortality. There is no specific treatment, though osseous lesions can regress spontaneously.

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