Reminder of important clinical lesson

A dilemma of fibrous dysplasia versus chronic osteomyelitis of the clavicle

Thisara Weerasuriya,¹ Sam Nashi,² Andrew Morgan,¹ Anele Ebizie¹¹Department of Orthopaedics, Tameside Hospital, Ashton-under-Lyne, Lancashire, UK²Liverpool Medical School, Liverpool, UK

Correspondence to Thisara Weerasuriya, drthisara@yahoo.co.uk

Summary

An 11-year-old child presented to the emergency department in 2005 with episodes of pain in his right clavicle. No history of trauma was noted and he was systemically well with no clinical signs of acute inflammation. He was extensively investigated and a diagnosis of fibrous dysplasia of the right clavicle was reached on histology. He developed periodic relapses of pain in the clavicle. He has been followed up since 2005, his most recent follow-up being in November 2011. The clinical dilemma of fibrous dysplasia versus chronic osteomyelitis is illustrated by highlighting this case.

BACKGROUND

The clinical dilemma of this case was to differentiate between chronic osteomyelitis and fibrous dysplasia of the bone. The latter is a rare genetic condition whereas the former is an infection. Differentiation between the two conditions is of paramount importance for treatment. This case highlights the diagnostic dilemma as well as the necessity of long-term follow-up of the patient as there is a less than 0.05% risk of malignancy associated with fibrous dysplasia of the bone. This case is of further interest as the latter condition is not commonly associated with the clavicle.

CASE PRESENTATION

An 11-year-old child presented to the emergency department in 2005 with a history of a painful bony lump in his right clavicle. There was no history of trauma and the child was systemically well at presentation. His right shoulder movements were full in range without any mechanical restriction. The neurovascular integrity of the ipsilateral upper limb was maintained.

X-rays of the clavicle demonstrated a bony thickening of the right clavicle at the junction of the middle and outer thirds of the clavicle, with sclerosis and a ground-glass smoky trabecular pattern (figure 1). Inflammatory markers were measured and the results of blood investigations were well within the normal range.

Differential diagnoses of chronic osteomyelitis, neoplasm, tuberculous osteomyelitis and fibrous dysplasia of the bone were made and further radiological imaging and bone biopsy were undertaken. A histological diagnosis of fibrous dysplasia of the bone was reached.

During the past 6 years the patient developed several episodes of recurrent pain in the right clavicle. Repeated blood investigations and imaging did not show evidence of inflammation or malignant changes.

INVESTIGATIONS

Inflammatory markers at presentation:

1. White cell count within a normal range for age with a normal neutrophil count.

2. C-reactive protein well within the normal range.
3. Erythrocyte sedimentation rate within the normal range for age.

X-rays of the right clavicle showed a widening of the junction of the middle and the lateral thirds of the right clavicle with a possible differential between chronic osteomyelitis and fibrous dysplasia of the clavicle being noted.

A nuclear bone scan showed an increased uptake of radioisotope in the region of the bony lump in the clavicle. Increased uptake was not noted at any other point in the rest of the skeleton.

On the basis of a CT scan of the clavicle, a differential between chronic osteomyelitis and fibrous dysplasia of the bone was considered (figure 2).

The MRI led to a differential between chronic osteomyelitis and fibrous dysplasia of the bone (figures 3–5).

Bone biopsy demonstrated a histological picture with misshapen bony trabeculae, with a fish-hook configuration and interspersed fibrous tissue. The trabeculae did not have a lining of osteoblasts. No neoplasm was noted. The histological diagnosis confirmed fibrous dysplasia of the bone (figure 6).

No acid-fast bacilli was noted on Ziehl-Neelsen staining.

Figure 1 X-ray of the right clavicle at presentation in 2005, showing thickening in the junction of the outer and middle thirds of the clavicle.

Figure 2 CT scan of the right clavicle (increased sclerosis with expansion).

DIFFERENTIAL DIAGNOSIS

- ▶ Chronic bacterial osteomyelitis
- ▶ Tuberculous osteomyelitis
- ▶ Fibrous dysplasia of bone
- ▶ SAPHO (synovitis, acne, pustulosis, hyperostosis, osteitis) syndrome
- ▶ Non-bacterial osteitis
- ▶ Bony neoplasm

TREATMENT

As the diagnosis was fibrous dysplasia of the bone the patient was reassured regarding the diagnosis and was followed up over the past 6 years. Long-term follow-up was necessary for two main reasons. First, fibrous dysplasia is

Figure 3 MRI scan of the clavicle showing cystic change, change of trabecular pattern and calcification.

Figure 4 MRI scan of the right clavicle.

known to be associated with malignant change with a risk of less than 0.05%; second, the patient had episodes of recurrent pain in the right clavicle.

OUTCOME AND FOLLOW-UP

The bony lump has been non-progressive over the past 6 years and repeat MRI scans have not demonstrated any malignant transformation. The risk of malignancy in fibrous dysplasia of the bone is less than 0.5%. Inflammatory markers have been repeatedly within normal range during the relapses of pain at the site of the bony lump of the clavicle.

DISCUSSION

The child's presentation to the department was with a painful lump in the clavicle without a history of trauma.

Figure 5 MRI scan of the right clavicle.

Figure 6 Histological image with misshapen bony trabeculae, with fish-hook configuration and interspersed fibrous tissue. The trabeculae did not have a lining of osteoblasts. No neoplasm was noted. The histological diagnosis confirmed fibrous dysplasia of the bone.

Otherwise he was systemically well and his inflammatory markers were within the normal range. The main issues in reaching a diagnosis were to exclude the possibility of chronic osteomyelitis and bony malignancy.

Imaging and the histology of the lump failed to show evidence of chronic osteomyelitis or bony malignancy, but showed a picture compatible with fibrous dysplasia of the clavicle.

Fibrous dysplasia of the bone is a benign condition.¹ It is progressive and is slow growing.¹

Fibrous dysplasia results from a defect in the differentiation of osteoblasts affecting maturation of the bone.¹ The condition is more common in the adolescent age group and our case was within the age group at presentation.

The sites of involvement are the femur (91%), tibia (81%), pelvis (78%), ribs, skull and facial bones (50%), upper limb, lumbar spine, clavicle and cervical spine in decreasing order of frequency.¹

Two thirds of patients are symptomatic before they reach 10 years of age.¹ Our patient presented at the age of 11 years and he did experience episodic pain during his 6-year follow-up period as well. The initial symptom is pain in the involved limb often associated with a limp or a pathological fracture, or both.¹ In our case, the patient had a lesion in the clavicle, which is not involved in direct weight bearing, and a pathological fracture was not involved in the presentation.

The structural integrity of the bone affected by fibrous dysplasia is weakened, and bowing of weight-bearing bones is common. Severe coxa vara deformity is known to occur in this condition when it affects the femur.¹

Establishing the diagnosis of fibrous dysplasia requires close cooperation between clinician, radiologist and pathologist. It is important to exclude other more serious conditions such as listed in the Differential diagnoses Section.

Radiographically, they are multiloculated lesions.² A sclerotic rim of bone is usually seen around these

lesions.² Pathological fracture is well described in larger lesions.²

Currently, there are no clearly defined systemic therapies for this bone disease.¹ Small, uncontrolled trials using second-generation bisphosphonates suggest that these bisphosphonates may be effective.¹ However, in the case discussed, as the clavicular lesion was relatively non-progressive and no pathological fracture was detected, no active treatment was commenced and the patient was followed up. The risk of malignancy is less than 0.5% in fibrous dysplasia of the bone.¹

SAPHO syndrome is a clinical syndrome first described by Khan *et al* in 1994, where the diagnostic criteria are multifocal osteomyelitis, sterile acute joint arthritis with or without pustular psoriasis, acne or hidradenitis, or sterile osteitis with skin manifestations.³ SAPHO syndrome was not considered as a possibility in our patient as there was no evidence of multifocal bone or joint involvement and no skin involvement was noted.

Non-bacterial osteitis is a sterile bone lesion. This can present with non-specific histopathological features of inflammation. The condition may be unifocal or multifocal. It is known to present acutely or chronically, and can be recurrent. Only when the condition is chronic, recurrent and multifocal, the term 'chronic recurrent multifocal osteomyelitis' is used for description.⁴ Our patient had specific histological features suggestive of fibrous dysplasia of the bone. The lesion was limited to the clavicle and there was no recurrence or appearance of multiple lesions so far during follow-up.

Learning points

- ▶ Think of common conditions first when encountered with a difficult clinical problem.
- ▶ Exclude the most sinister causes listed in the differential diagnosis.
- ▶ Confirm the diagnosis with the appropriate investigation.
- ▶ Investigations are aides to clinical judgement.

Competing interests None.

Patient consent Obtained.

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