Management of benign bone tumours

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Abstract

Benign bone tumours are rare, occurring most commonly in skeletally immature patients, and usually arising from cartilage or bone. The commonest locations are the distal femur, proximal tibia and proximal humerus. They present with pain, swelling or pathological fracture. Diagnosis is by plain radiographs, MRI and core needle biopsy if indicated. More aggressive tumours may appear radiologically similar to malignant tumours. Management depends upon anatomical location, symptoms, morbidity of treatment and the natural history of the tumour. In most cases it involves either simple excision or curettage, although occasionally it is necessary to perform a complete excision using the same principles as for malignant tumours.

Keywords benign bone tumour; non-neoplastic tumour-like conditions of bone

Introduction

Primary bone neoplasms are extremely rare, accounting for only 0.2% of human tumours.[1] The majority are benign and typically affect the skeletally immature patient but some are difficult to distinguish from malignant lesions, have a significant incidence of local recurrence and may undergo malignant transformation. Diagnosis and treatment of bone tumours is complex and management of this group of patients is best undertaken at specialist centres in a multidisciplinary (MDT) setting.

Classification

Two classification systems are commonly used (Tables 1 and 2). The first is histological, based on the cell of origin. The second is more clinically orientated and based on the pattern of behaviour of the tumour.[2]

While tumours may arise from chondrocytes, osteoblasts, osteoclasts, or soft tissue within bone such as fat, fibrous tissue or smooth muscle, some fall into the 'tumours of undefined neoplastic nature' group and their pathogenesis is unclear. Their behaviour dictates their clinical presentation, varying from being an incidental finding on radiography in the case of a latent lesion, to a rapidly growing, painful lesion associated with functional loss in the case of an aggressive lesion. This is reflected in Enneking's classification (Table 2).[2]

Classification of tumour by histological subtype

Cell type	Tumour subtype
Chondrogenic	Osteochondroma
	Chondromas: enchondromas, periosteal chondroma
	Chondroblastoma
	Chondromyxoid fibroma
	Bizarre parosteal osteochondromatous proliferation
Osteogenic	Osteoma (enostosis/bone island)
	Osteoid osteoma
	Osteoblastoma
Fibrogenic	Desmoplastic fibroma of bone
Fibrohistiocytic	Non-ossifying fibroma
	Fibrous cortical defect
Osteoclastic	Giant cell tumour
Vascular	Haemangioma
Lipogenic	Intra-osseous lipoma
Tumours of undefined neoplastic nature	Aneurysmal bone cyst
	Simple bone cyst
	Fibrous dysplasia
	Osteofibrous dysplasia

Table 1

Aetiology

The aetiology of the vast majority of benign bone tumours is unclear. Numerous theories have been proposed but none has ever been substantiated apart from multiple hereditary exostoses (MHE), which is also known as diaphyseal aclasis. MHE is inherited as an autosomal dominant condition. Three genes are responsible, namely the exostosin-1/2/3 (EXT-1,2,3) genes found at chromosome 8q24 (EXT-1) loci, chromosome 11p11-12 (EXT-2) loci and the short arm of chromosome 19 (EXT-3), although its exact location has yet to be precisely determined. Approximately 15% of patients with osteochondromas have the inherited form of the condition.

Clinical features

The clinical features are non-specific and variable. Some long-standing lesions present as incidental findings. For example, an adolescent who attends the A&E department after a knee injury and a fibrous cortical defect is found on radiography (Figure 1). Some tumours such as osteochondromas present with a long history of a painless swelling, but more aggressive lesions such as giant cell tumours (GCT) usually present with a short history of pain, swelling and loss of function.

Investigation

As some benign bone tumours can be difficult to distinguish from malignant lesions, thorough and well-considered investigation

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Classification of tumour by biological behaviour according to Enneking[2]

Classification	Behaviour	Example of tumour
Latent	Slow growth with spontaneous healing. Often an incidental finding on X-ray. No treatment required.	Fibrous cortical defect Non-ossifying fibroma Osteoma
Active	Progressive growth over time and usually symptomatic. Treatment of choice; curettage. Low incidence of local recurrence.	Chondromyxoid fibroma Enchondroma Simple bone cyst
Aggressive	Rapid growth of tumour often extending beyond periosteum into the soft tissues. Treatment of choice curettage or excision. 10–15% chance of local recurrence	Chondroblastoma osteoblastoma Giant cell tumour aneurysmal bone cyst

Table 2

should be undertaken in the MDT setting, in a specialist musculoskeletal tumour centre.[3] The absolute minimum includes an adequate history, examination, and imaging by radiography, MRI and occasionally CT and ultrasound.

The radiological features may be so clear that if compatible with the clinical picture, a histological diagnosis may not be necessary prior to definitive treatment. However, if there is any doubt about the diagnosis or worrying features on imaging or examination, a tissue diagnosis should be obtained. This is best achieved by image-guided percutaneous bone biopsy using a Jamshidi™ needle, with the biopsy route agreed between the surgeon and radiologist at the MDT.

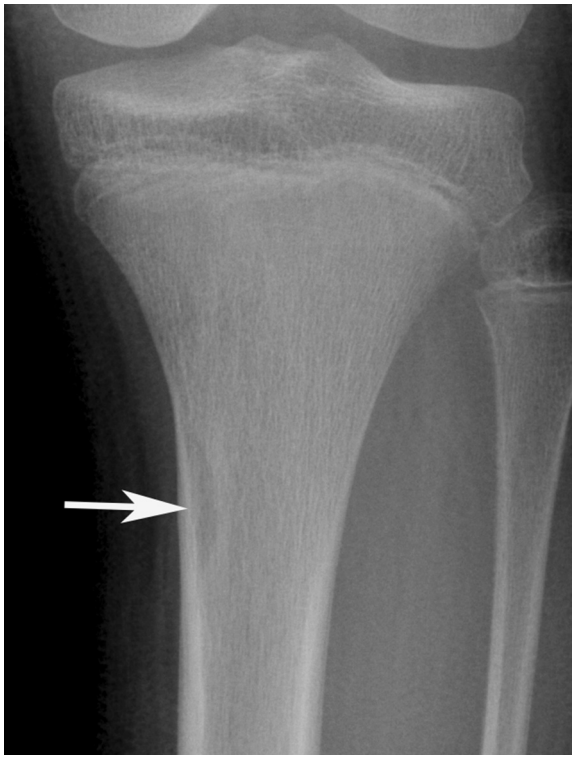

Figure 1 Antero-posterior radiograph of the proximal tibia showing a small fibrous cortical defect (arrow) in the medial tibial metaphyseal cortex.

Management

General principles

Treatment is dependent upon many factors, particularly:

- patient symptoms
- natural history of the tumour
- morbidity of treatment.

Treatment varies from simple observation with repeat imaging, through to wide excision using the same surgical principles as for malignant tumours. When treating bone tumours, the surgeon has to balance excision margin against function. With wider margins, there may be greater functional loss but a reduced risk of local recurrence. Conversely, intra-lesional surgery has less morbidity but a greater risk of local recurrence.

Non-operative

Asymptomatic lesions that fall into the Enneking latent group of tumours can simply be observed, for example the natural history of lesions such as non-ossifying fibromas is well-documented and predictable, and it is safe to leave these alone. If the diagnosis has been made on imaging alone and non-operative treatment has been chosen, then histological confirmation of the diagnosis will not be available. Therefore, it is advisable to repeat the plain radiograph after 3–6 months to ensure that the lesion is showing no signs of progression. If clinical or radiological progression does occur, then there must be a low threshold for biopsy.

Curettage

Curettage is the treatment of choice for the majority of benign bone tumours requiring surgical intervention, and is by definition intra-lesional surgery. The intention is to achieve macroscopically clear margins, accepting that microscopic disease is likely to be left behind.

The technique involves exposing the affected bone and creating a bone window with osteotomes. The window needs to be big enough to obtain an adequate view of the tumour but small enough to ensure that the graft material can be contained within the bone at the end of the procedure. Ideally, a range of curettes with differing angles and head sizes should be used. In addition, it is now standard practice to skim the edge of the cavity with a high-speed burr to reduce the risk of local

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