Osteoid osteoma and benign osteoblastoma in childhood

J. A. BLACK, R. K. LEVICK, AND W. J. W. SHARRARD

The Children's Hospital, Sheffield

SUMMARY Three cases of osteoid osteoma and one of benign osteoblastoma in children are described. The main complaint was severe pain which was worse at night; it was relieved by aspirin or other analgesics. The diagnosis was made on clinical and radiological grounds and was confirmed on histological examination of the central nidus removed at operation. The pain was relieved in the patients with osteoid osteoma, and it was very much less after operative removal of the benign osteoblastoma. Both conditions are probably variations of the same disease process, depending on the anatomical site and the type of bone affected.

Osteoid osteoma was described as a clinical entity in 1935 by Jaffe, and then more fully by Jaffe and Lichtenstein (1940). In 1932 Jaffe and Mayer reported a patient in whom the disease differed somewhat from osteoid osteoma, and Dahlin and Johnson (1954) further described such cases and used the term 'giant osteoid osteoma'. The term benign osteoblastoma was introduced by Jaffe (1956) and Lichtenstein (1956) independently to differentiate this condition from osteoid ostemal. Since then numerous reports on both conditions have appeared, mainly in orthopaedic and radiology journals. In our experience children with these conditions are referred about equally to general paediatric and orthopaedic clinics. Nevertheless the clinical picture, which is similar in both osteoid osteoma and benign osteoblastoma, appears to be unfamiliar to many paediatricians. It was for this reason, and also because of the inherent difficulty in making the diagnosis, that we describe our experience with three patients with osteoid osteoma and one with benign osteoblastoma seen in Sheffield during the last 12 years.

Case reports

Case 1. Osteoid osteoma, left femur. This boy was born on 2 October 1958 and was first seen at the orthopaedic clinic at age 7½ years with a history of pain in the left thigh and a limp for a year. The pain was not constant and was relieved by aspirin. An x-ray showed irregular sclerosis of the medial side of the upper half of the shaft of the left femur; there was a faint oval translucent area at the level of the lesser trochanter, which was confirmed on tomography. A radiological diagnosis of osteoid osteoma was made (Fig. 1).

Fig. 1 Case 1. Osteoid osteoma: sclerosis of medial side of femoral shaft and faint oval translucency at level of lesser trochanter.
A small piece of bone was removed from the upper part of the femur but this did not show any histological evidence of osteoid osteoma. The pain was relieved for a short time but returned when he started walking. Two months later, further x-rays showed that the lucent area was still present; the pain was now very severe. At age 8 years he was readmitted for observation but did not appear to have any pain. One month later he was having to take 'up to 3 aspirins' daily. Further x-rays and tomography again suggested osteoid osteoma, and the femur was explored; this was nearly one year after his first visit to the hospital and 2 years after the onset of symptoms. On this occasion several fragments of bone were removed. One of these contained a nidus of irregular, partially calcified osteoid trabeculae separated by loose vascular connective tissue containing osteoblasts and osteoclasts, and bounded by dense lamellar bone. The appearance was consistent with the clinical diagnosis of osteoid osteoma. After this operation the boy was free from pain.

He remained well until October 1973, when at age 15 years, he complained of being always tired; he was very pale. Hb was 7.6 g/dl and blood film showed pronounced iron deficiency anaemia. He admitted that he had taken aspirin since his original trouble and that he was now taking it for headaches. His urine showed a faint purple coloration with Phenistix, and plasma salicylate level was 6 mg/100 ml (0.43 mmol/l). He was readmitted (J.A.B.) for further investigation. Apart from extreme pallor, no abnormality was found except that his stools showed a positive result in a test for occult blood. Barium swallow and follow-through radiological examination were normal. He was given ferrous sulphate tablets and returned home. Six weeks later Hb was 11 g/dl; the ferrous sulphate was finally stopped 3 weeks later when Hb was 13.8 g/dl. He felt well; there was no wasting of the thigh, and knee and ankle jerks were normal on both sides.

This case shows how difficult it is to localise the lesion at operation, in spite of clear radiological evidence of its existence. Aspirin dependence is a hazard to which people with osteoid osteoma are particularly prone, and there is no doubt that taking aspirin regularly for a long period caused severe iron deficiency anaemia from gastrointestinal bleeding.

**Case 2. Osteoid osteoma, right femur.** This boy was born on 18 March 1962 and referred to one of us (J.A.B.) at age 10 years with a 4-month history of pain in the right leg. The pain was not continuously severe but was worse at night, waking him at about 2 a.m.; it was relieved by aspirin and he had been taking one tablet each night for the previous 2 months. He had also been noted to limp occasionally and for one month his right leg had been getting thinner.

There was 2.5 cm of wasting of the muscles in the right midthigh, and knee and ankle jerks were reduced on the right side; there was also slight wasting of the right gluteal muscles. No muscle weakness or sensory loss was detected. A lumbar puncture showed no evidence of block, and the CSF was normal. The only radiological abnormality was a thickening of the cortex at the level of the lesser trochanter (Fig. 2).

He was referred to the orthopaedic department (W.J.W.S.); there was some tenderness of the upper half of the right femur and further x-rays of the femur with different degrees of rotation confirmed the cortical thickening in the upper half of the shaft; there was no evidence of a lucent central lesion after tomography, but osteoid osteoma was considered likely. He was readmitted to hospital and a block of bone at the site of the cortical thickening was removed. The tissue contained a central lesion which was histologically similar to that described in Case 1 and was typical of an osteoid osteoma; his pain was completely relieved. The total duration between the onset of symptoms and operation was 8 months. 10 months after the operation he was free...
of symptoms; there was no wasting of the thigh, and his knee and ankle jerks on the right side were normal.

The wasting and reduced reflexes at first suggested a neurological lesion, but the radiological evidence strongly suggested an osteoid osteoma although alucent area was never detected by radiography.

Case 3. Osteoid osteoma, left tibia. This girl was born on 24 October 1964 and referred at age 41/2 years. She complained of a cramp-like pain in the left leg and a limp for the previous 2 months. The pain was particularly severe at night and was relieved by aspirin. There was slight wasting (2 cm) of the left thigh and calf muscles, with diminished muscle power in the left leg. The left ankle jerk was slightly diminished compared with the right side. X-rays showed cortical thickening of the posterolateral aspect of the lower tibial shaft (Fig. 3). There was a small lucent area which appeared to be deep to the sclerosis, and the diagnosis of osteoid osteoma was suggested.

She was admitted in May 1969 and at operation, using radio-opaque markers, a typical osteoid osteoma was removed; the diagnosis was confirmed histologically. The pain was completely relieved.

Case 4. Benign osteoblastoma, right ischium. This boy was born on 4 December 1958 and, apart from lobar pneumonia at age 11 years, he had been well until he developed an aching pain in his right upper thigh at 13 years. He was seen at a paediatric out-patient clinic (J.A.B.) and was found to have 2 cm of wasting of the right thigh and 1·5 cm of wasting of the calf, with diminished knee and ankle jerks on the right side; there was a doubtful area of reduced sensation to pin pricks on the front of the right thigh.

Because of the possibility of compression of the right femoral nerve an IV urogram was done which showed an indentation of the bladder by a soft-tissue mass on the right side of the pelvis, and some sclerosis and thickening of the right ischium (Fig. 4). The ESR was 21 and 48 mm/in 1st hour. The right side of the pelvis was explored by an anterior approach; the right levator ani was bulkier than normal. A small piece of bone removed at operation showed no abnormality. The pain remained unchanged and he started taking paracetamol to relieve it. After 6 months' observation as an out-patient there was no improvement and he became a thin worried boy. He was referred to orthopaedic out-patients department (W.J.W.S.) where further x-rays, including tomograms, showed an area (1·5 cm in diameter) of lucency in the upper part of the right ischium. A Brodie's abscess was considered probable. He was readmitted and at exploration of the ischium an area of soft bone was scooped out. The histological appearance of the tissue removed was similar to that described in Case 1, except that the connective tissue separating the osteoid trabeculae was somewhat less vascular.

After operation his pain was almost completely relieved. He was last seen in 1973 (aged 14-8 years) when he still complained of slight pain: there was now only 0·6 cm of wasting of the right thigh, and knee and ankle jerks were equal on both sides. He was a cheerful boy and had gained 8 kg since the operation.

Discussion

Osteoid osteoma is twice as common in boys as in girls, with a maximum age incidence of between 10 and 25 years, and extremes from 1½ and 60 years (Poulsen, 1969). The presenting symptom is pain which is sometimes intermittent at first but later becomes continuous. The pain may vary in intensity and is characteristically worse at night. It has been
described as aching or boring, and patients usually obtain relief from such drugs as aspirin; this may cause dependence upon them (Case 1). Localisation is usually easy if a long bone is affected, but difficult if the lesion is in the upper part of the femur, the pelvis, or a vertebra. In order of frequency, the tibia and femur are most commonly involved, followed by the tarsal phalanges and the vertebrae, but almost any bone can be affected with the possible exception of the maxilla and facial bones. Lesions in the mandible (Flaherty et al., 1956) and skull (Munk et al., 1960) are rare. Epiphyseal lesions are rare but Sherman (1947b) described a patient with a lesion in the epiphysis of the great trochanter. Occasionally the pain is referred from the lumbar spine to the abdomen or leg, from the lesser trochanter to the knee, or proximally from the tibia to the hip (Golding, 1954), or thigh (Vaughan-Jackson, 1950). Very rarely the lesion is painless and presents as a swelling (Lawrie et al., 1970). Other symptoms depend on the site of the lesion; if the shaft of the femur or tibia is affected there may be a limp, with wasting of muscles of the thigh, accompanied by loss or depression of knee and ankle jerks (Cases 1, 2, and 3). Rushton et al. (1955) found a positive Laségue's sign in two cases with lesions of the femoral shaft, and one patient with loss of sensation to pain and touch over the anterolateral part of the thigh. Vertebral lesions may cause scoliosis, particularly in children and adolescents (Freiberger, 1960; Poulson, 1969). In lesions near the elbow joint there is loss of flexion and extension with preservation of pronation and supination, and osteomyelitis may be suspected (Marcove and Freiberger, 1960). Szepesi et al. (1972) described two children with lesions in the great trochanter and femoral neck who had flexion contractures at the hip joints, and Johnson (1955) reported two patients with intracapsular lesions of the femoral neck in whom there was a chronic synovitis with fluid in the joints and infiltration of the synovial membranes with round cells. In young children deformity caused by uneven bone growth may develop if the lesion is near an epiphyseal plate (Ponseti and Barta, 1947), and in some cases the affected limb is longer than the normal one. Physical signs are varied; wasting rarely occurs except in the thigh or calf but a clinically evident effusion into an adjacent joint has been described (Sherman, 1947a; Spence and Lloyd-Roberts, 1961) and, particularly at the wrist and ankle, a nearby bone may show hyperostosis (Morton and Bartlett, 1966) or osteoporosis. Locally there may be exquisite tenderness and a slight increase in temperature. In areas easily accessible to palpation, thickening of the bone may be detectable. When a terminal phalanx is affected the whole finger end is swollen and dusky red, with excessive sweating and hypertrophy of the nail, and premature fusion of the phalangeal epiphyses has been observed (Rosborough, 1966). A lesion in a metatarsal bone may be mistaken for a march fracture (Kallio, 1963).

The clinical diagnosis of osteoid osteoma is
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Notoriously difficult; the interval between the onset of symptoms and a correct diagnosis is seldom less than 6 months and it may be many years; it is not surprising, particularly in adults, that the depression, insomnia, and loss of weight produced by the long-continued pain often result in a diagnosis of hysteria or psychoneurosis. A limp, with wasting and reduced, or absent, knee and ankle jerks, may suggest compression of the femoral nerve or lumbosacral plexus, a cord lesion, or protrusion of an intervertebral disc. Brodie's abscess (Szepesi et al., 1972) may be considered on clinical and radiological grounds, particularly if the sedimentation rate is slightly raised, which can occur in osteoid osteoma. O'Hara et al. (1975) considered that Brodie's abscess and stress fracture of the tibia were likely to cause the greatest diagnostic difficulties. Other conditions which may have to be considered (Purcell et al., 1952) are solitary cortical abscess of bone, chronic sclerosing osteomyelitis of Garré, solitary tuberculous lesion in the shaft of a long bone, Ewing's tumour or osteogenic sarcoma, and chronic synovitis in cases with an effusion into an adjacent joint. A central cavity does not occur in chronic sclerosing osteomyelitis, but is found in a chronic tuberculous lesion. In case of doubt culture of material from the central lesion and microscopical examination should settle the diagnosis. The two malignant bone tumours can usually be distinguished by their rapid progression and destruction of bone. Lindbom et al. (1960) and O'Hara et al. (1975) have shown by angiography that the central area of an osteoid osteoma is extremely vascular, while a Brodie's abscess has an avascular centre.

Radiologically the lesion can usually be seen as a well-defined area of dense sclerosis, with a diameter of 1/2-1 cm, occasionally up to 2 cm. In the centre of the sclerosis is a lucent nidus, sometimes containing speckled areas of calcification which may be mistaken for sequestra (Munk et al., 1960).

Tomography may be necessary to demonstrate the central lesion, particularly if there is a considerable amount of thickened sclerotic bone surrounding it and, in some cases, a central area cannot be defined at all (Case 2). If the lesion is near the surface of the cortex or just under the periosteum, the long bones, especially the cortical bone, will be much thickened. Conversely, thickening is less pronounced if the lesion is in spongy bone. The thickened cortical bone can easily be missed unless a number of views are taken with different degrees of rotation (Case 2). Purcell et al. (1952) pointed out that thickened layers of subperiosteal bone may produce an 'onion peel' appearance similar to that seen in Ewing's tumour.

Spontaneous cure may occur after many years (Vickers et al., 1959) and increased calcification in the central area has been regarded as evidence of healing (Moberg, 1951; Golding, 1954). However, the only effective treatment is complete excision of the whole central lesion. Accurate localisation of the lucent central area at operation is often difficult and may require the placing of markers under radiological control. In most instances the removal of a block of bone surrounding the actual lesion is preferable to an attempt to shell out the nidus as a small piece of tissue may be left behind; packing with an autogenous bone graft is sometimes required. Incomplete removal is followed by a recurrence of pain which can be relieved only by a further operation. Other methods of treatment, including radiotherapy, are useless.

Histologically the central lesion consists of an area of highly vascular tissue with wide vascular spaces and thick-walled blood vessels. There is much osteoid tissue, in irregular seams, with numerous osteoblasts. Unmyelinated nerve fibres have been found in the central area (Chandler and Kaell, 1950; Sherman and McFarland, 1965; Byers, 1968; Schulman and Dorfman, 1970), mainly in relation to the blood vessels. The aetiology of osteoid osteoma is obscure. Trauma seems to play a part in some cases (Marcove and Freiberger, 1960; Rosborough, 1966), particularly in the phalanges. The cause of the pain is not fully understood, but Golding (1954) showed that the pain in a metacarpal lesion could be relieved by applying a pneumatic tourniquet, inflated above the systolic pressure, to the upper arm. Sherman and McFarland (1965) pointed out that bone itself is relatively insensitive to pain and that bone lesions normally cause pain only when the periosteum is affected. Osteoid osteoma, however, is painful right from the start, irrespective of its anatomical relationship to the periosteum. Sherman and McFarland suggested that the unmyelinated fibres found in the lesions are autonomic nerves which are stimulated by distension of the blood vessels, causing pain and also the vascular dilatation and sweating which have sometimes been noted (Chandler and Kaell, 1960; Rosborough, 1966). The cause of the muscle wasting and depression of reflexes is difficult to explain as the weakness of the affected muscles is slight and certainly not sufficient to affect the reflexes: nor in most cases is there any significant amount of disuse of the affected limb. The vascular symptoms are similar to those found in glomus tumours (Kohout and Stout, 1961; Laymon and Peterson, 1965) in which there is sometimes vasodilatation and increased sweating of an affected finger (Stabins et al., 1937) similar to that seen in a few cases of osteoid osteoma.

Benign osteoblastoma was differentiated from
osteoid osteoma by Dahlin and Johnson (1954) who at first used the term giant osteoid osteoma. In 1964 Lichtenstein and Sawyer reviewed reports and added a number of new cases. The age, sex incidence, and distribution of lesions in benign osteoblastoma are similar to those of osteoid osteoma, but the former occurs more often in spongy than in cortical bone. Pain is less severe than in osteoid osteoma and is less easily located. The lesion is generally larger (2.5–5 cm in diameter), less regular in shape, and there is more thickening of the surrounding soft tissue (Case 4). When a vertebra is affected there may be compression of the cord or roots (Lichtenstein and Sawyer, 1964). Radiologically, there is an expanding area of bone destruction, usually with a sclerotic margin; when the surrounding bone is perforated a soft tissue mass is formed which may be identifiable in x-rays (Case 4). The differential diagnosis is similar to that of osteoid osteoma but should also include giant-cell tumour, fibrous dysplasia, aneurysmal bone cyst, nonossifying bone fibroma, and eosinophil granuloma (Wellmer et al., 1968). Angiography may be helpful in difficult cases, as in osteoid osteoma. Treatment is by excision. In benign osteoblastoma the osteoid tissue is less regularly arranged than in osteoid osteoma and there are more multinucleated cells in addition to the osteoblasts. It remains uncertain, however, whether the two conditions are completely separate or are variations of the same disease process. A third, apparently related condition with multiple lesions similar to osteoid osteoma has also been described (Schajowicz and Lemos, 1970). De Souza Dias and Frost (1974) regard all three conditions as osteoblastoma and suggest a classification according to site: cortical osteoblastoma, spongy osteoblastoma, periosteal osteoblastoma, and multifocal osteoblastoma. They considered that the radiological appearance depended on the site of the lesion; in compact bone or near cortex (osteoid osteoma) there was considerable tissue reaction, while in spongy bone (osteoblastoma) there was relatively little reaction. Schajowicz and Lemos (1970) also pointed out that an insistence that these lesions are always benign is probably unjustified in our present state of knowledge.

We thank Professor J. L. Emery and Dr Gillian Batcup of the Department of Histopathology, Children’s Hospital, Sheffield, for the reports on the sections.

References


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Correspondence to Dr J. A. Black, Department of Paediatrics, The Children’s Hospital, Western Bank, Sheffield S10 2TH, Yorkshire.

Received 8 August 1978

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J A Black, R K Levick and W J Sharrard

Arch Dis Child 1979 54: 459-464
doi: 10.1136/adc.54.6.459

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