Neurological manifestations of osteoid osteoma

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Abstract
The clinical and radiological features of 38 children with osteoid osteomas were analysed retrospectively. Twenty nine patients had lesions of the femur (n=17) or tibia (n=12). The mean duration from the onset of symptoms to diagnosis was 13·8 months. In seven patients the history of pain and abnormalities on examination suggested a possible neurological disorder. Fourteen of 29 patients (48%) with femoral or tibial osteomas had localised muscle atrophy, and 10 patients (34%) had diminished or absent deep tendon reflexes in the affected limb. Two patients had painless lesions. Six patients had normal plain radiographs. Delay in the diagnosis of osteoid osteoma may be prevented by the knowledge that pain may be referred or radicular, that the concomitant occurrence of muscle atrophy and depressed deep tendon reflexes are relatively common findings, and that the characteristic radiological features may only appear late in the course of the disease.

Osteoid osteoma is a benign tumour of bone. Orthopaedic surgeons are well acquainted with the lesion, which characteristically produces intense pain (initially nocturnal) and relieved by salicylates.¹ Symptoms are often present for many months before the correct diagnosis is made, partly because the characteristic radiological findings may not appear until late in the course of the disease.² In addition, the tumour may mimic certain neurological diseases, particularly spinal lesions, thereby resulting in unnecessary neuroradiological investigations.³ Because of the varied presentation of osteoid osteoma and the inherent difficulty in making the diagnosis, we have analysed systematically

the clinical and radiological features of this lesion, with particular reference to the signs and symptoms suggestive of neurological disorder.

Patients, methods, and results
The records of the Royal Children's Hospital from 1975–89 were searched for patients with a diagnosis of osteoid osteoma. Thirty eight were identified, in 36 of whom there was histological confirmation of the diagnosis. The latter two patients (cases 14 and 35) had typical clinical histories of deep nocturnal pain that was relieved by salicylates, and characteristic changes on plain radiographs and computed tomograms. All patients presenting with neurological symptoms had been examined by a paediatric neurologist. There were 24 boys and 14 girls (ratio 1·7:1) with a mean age of 9·3 years (range 3–17 years). Twenty nine patients (76%) had lesions of the femur (n=17) or tibia (n=12). Of the remaining patients, lesions were located in the proximal humerus (n=3), phalanges (n=2), vertebrae (n=2), ulna (n=1), and calcaneus (n=1). The mean duration from the onset of symptoms to diagnosis was 13·8 months (range 2–60 months).

CLINICAL FEATURES
On the basis of the clinical features, mode of presentation, and site of the lesion, the patients were divided into four groups.

Group 1
Seven patients were initially considered to have neurological problems. The presenting symptoms were predominantly disturbance of gait and muscle wasting, with pain being a less

Table 1 Clinical features of patients in group 1

Case No	Age (years)	Sex	Site of lesion	Duration of symptoms (months)	Presenting features	Findings on examination
1	4	M	Femur	2	Leg wasting; limp	Wasted right quadriceps and gastrocnemius; 4/5 weakness knee extension; absent right knee jerk
2	6	M	Femur	12	Leg wasting; limp; thigh pain	Wasted left quadriceps; 4/5 weakness hip flexion and knee extension; absent left knee jerk
3	17	F	Femur	18	Leg wasting; anterior thigh pain	Wasted left quadriceps; absent left knee jerk
4	8	F	Femur	30	Leg wasting; limp; anterior thigh pain	Wasted left quadriceps, gastrocnemius, and glutei; depressed left knee and ankle jerk
5	10	M	Femur	6	Limp; anterior thigh pain; testicular pain	Wasted left quadriceps; depressed left knee jerk
6	8	F	Femur	2	Limp; thigh calf, and shin pain; leg wasting	Wasted right quadriceps and gastrocnemius; depressed right knee and ankle jerk
7	6	M	Tibia	12	Limp; anterior leg pain; leg wasting	Wasting and 4/5 weakness right tibialis anterior, hamstrings, gastrocnemius, and glutei; absent right ankle jerk

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prominent feature. Six patients had femoral osteomas and one had a tibial osteoma (table 1). Four patients had normal radiographs of the lumbosacral spine and normal myelograms before the correct diagnosis was made. All patients had been initially referred to a neurologist.

Group 2

Seven patients presented with orthopaedic problems, with hip or leg pain, but on examination were found to have localised muscle wasting or depressed deep tendon reflexes, or both, in the affected limb (table 2).

Group 3

Fifteen patients, six with femoral osteomas and

nine with tibial osteomas, presented without neurological symptoms or signs. All except one patient presented with either local or referred pain (table 3).

Group 4

Nine patients had osteoid osteomas in sites other than the femur or tibia. One patient with an osteoma in the proximal humerus presented with a palpable bony swelling resulting in brachial plexus compression. The remaining patients presented with localised pain (table 4).

Although there was considerable variation in their clinical features, all patients except two experienced varying degrees of pain. Case 1 presented with a limp and leg wasting without pain or tenderness, and case 21 presented with a

Table 2 Clinical features of patients in group 2

Case No	Age (years)	Sex	Site of lesion	Duration of symptoms (months)	Presenting features	Findings on examination
8	6	F	Femur	12	Thigh and shin pain	Wasted left quadriceps, gastrocnemius; 4/5 weakness knee extension; depressed left knee and ankle jerk
9	4	M	Femur	12	Thigh and leg pain	Wasted left quadriceps; limp
10	8	M	Femur	60	Thigh pain	Wasted left quadriceps; 1 cm discrepancy in leg length
11	12	M	Femur	24	Knee pain	Wasted left quadriceps and gastrocnemius; depressed left knee jerk
12	10	F	Femur	12	Knee pain	Wasted left quadriceps; depressed left knee jerk, reduced hip flexion and abduction
13	14	M	Tibia	3	Leg pain	Wasted right gastrocnemius, 4/5 weakness in plantar flexion
14	9	M	Tibia	30	Leg pain, and bony swelling	Wasted left quadriceps; bony swelling left tibia

Table 3 Clinical features of patients in group 3

Case No	Age (years)	Sex	Site of lesion	Duration of symptoms (months)	Presenting features	Findings on examination
15	13	M	Femur	4	Hip pain	Not recorded
16	9	M	Femur	18	Knee and groin pain	Not recorded
17	12	F	Femur	60	Thigh pain	Not recorded
18	10	M	Femur	2	Hip and knee pain	Reduced flexion and abduction left hip
19	10	M	Femur	6	Hip pain	Reduced range of movement right hip
20	3	M	Femur	6	Thigh pain; limp	Local tenderness
21	13	M	Tibia	18	Painless swelling, left tibia	Bony swelling medial aspect left tibia
22	14	M	Tibia	Not known	Swelling left tibia, leg pain	Bony swelling proximal left tibia
23	9	M	Tibia	Not known	Leg pain	Not recorded
24	8	M	Tibia	2	Leg pain, swelling left tibia	Bony swelling left tibia
25	6	M	Tibia	18	Ankle pain	Bony swelling distal right tibia
26	11	M	Tibia	3	Leg pain; limp	Not recorded
27	11	F	Tibia	12	Knee pain	Bony swelling proximal right tibia
28	9	F	Tibia	2	Leg and ankle pain; limp	Not recorded
29	4	M	Tibia	6	Leg pain; limp	Bony swelling right tibia

Table 4 Clinical features of patients in group 4

Case No	Age (years)	Sex	Site of lesion	Duration of symptoms (months)	Presenting features	Findings on examination
30	10	F	Humerus	18	Elbow pain	Tender swelling distal right humerus; reduced elbow extension
31	13	F	Humerus	12	Arm pain	Tender swelling upper medial humerus; wasting and weakness C5, 6, 7 and sensory loss C5 and 6
32	14	F	Humerus	Not known	Arm pain	Not recorded
33	6	F	Proximal phalanx right fifth finger	Not known	Painful swelling fifth finger	Tender swelling right fifth finger
34	9	M	Distal phalanx second toe	Not known	Pain second toe	Not recorded
35	11	M	L5 vertebra	5	Low back pain (nocturnal)	Not recorded
36	13	M	S2 vertebra	12	Limp; left thigh and buttock pain	Wasting left hamstrings and calf; weakness left hamstrings, and quadriceps, positive Trendelenburg's test
37	7	M	Ulna	Not known	Pain left forearm	Tender swelling left ulna
38	16	F	Calcaneus	12	Pain left foot	Tender swelling lateral aspect left foot

painless swelling of the proximal tibia. Pain was usually felt in the immediate region of the bone lesion, but many patients had referred pain, usually from a femoral osteoma to the knee. Some patients experienced pain in regions remote from the lesion, without local pain. Case 5 presented with left testicular pain, and later pain in the left anterior thigh, from a lesion in the left femoral neck. Case 6 had posterior thigh, calf, and shin pain from an osteoma in the femoral neck. Cases 11 and 12 experienced only knee pain, and case 16 had knee and groin pain from femoral osteomas. Case 36 complained of radicular pain in the buttock and posterior thigh from an osteoma in the left neural arch of S2.

Atrophy of some muscles of the affected limb was noted in 14 (48%) of 29 patients with femoral or tibial osteomas. One patient (case 36) with an osteoma in the left neural arch of S2 had wasting of the left hamstring and calf muscles. Those muscles in the immediate region of the lesion were most commonly affected. In five patients with femoral osteomas, however, calf wasting was present in addition to wasting of the thigh (fig 1). One patient with a tibial osteoma had quadriceps as well as calf wasting

Figure 1 Case 1: wasting of right vastus lateralis, vastus medialis, and gastrocnemius in a patient with an osteoid osteoma of the right femoral neck.

(case 14), and one patient had wasting of the buttock and hamstrings (case 7). Muscle weakness was uncommon despite the presence of muscle wasting. Two patients in group 1 (neurological presentation) had mild quadriceps weakness, and one patient (case 7) with a tibial osteoma had mild weakness of tibialis anterior and gastrocnemius. One patient in group 2 (case 8) had mild weakness of quadriceps. Activity of the deep tendon reflexes of the affected limb was diminished or absent in 10 (34%) of 29 patients with femoral or tibial osteomas. All seven patients in group 1, who were initially thought to have neurological problems, had depressed or absent reflexes in the affected limb.

Twelve patients limped on the affected lower limb, the diagnoses being femoral osteoma (n=5), tibial osteoma (n=6), and sacral osteoma (n=1). The limp was usually attributed to pain, although in three patients mild muscle weakness was also present. Local swelling, usually associated with tenderness, was present in eight (67%) of 12 patients with tibial osteomas, two with humeral osteomas, and in single patients with lesions in the ulna, calcaneus, and proximal phalanx of the fifth finger. No patient had sensory loss on examination with the exception of case 31, in whom it was caused by compression of the brachial plexus by the bony swelling.

IMAGING STUDIES

All patients had plain radiographs of the relevant region, of which 30 (79%) showed findings strongly suggestive of osteoid osteoma. One patient (case 19) had findings that were abnormal but equivocal (not being diagnostic of osteoid osteoma), and in one patient with an osteoma of the femoral neck (case 3), radiographs showed only subluxation of the hip joint. Six patients had normal plain radiographs of the relevant region of which one had an abnormal tomogram. Five of the six patients had femoral neck osteomas, and one had a vertebral osteoma. In one patient, abnormalities were noted on a repeat plain radiograph six months after presentation.

In 17 patients radionuclide scans were carried out. Ten were positive, showing intense, well defined, focal areas of increased activity on the radioangiogram and blood pool studies. Seven were abnormal but not diagnostic of osteoid osteoma. Fifteen patients had computed tomograms, all of which were abnormal with findings strongly suggestive of osteoid osteoma. One patient did not undergo surgical excision of the lesion, as the parents withheld consent to operation. The initial plain radiographs and computed tomogram showed the characteristic appearances of an intracortical osteoid osteoma. Repeat imaging studies 15 months later showed obvious healing of the lesion with only a small area of sclerosis on the plain radiograph, and normal bone scan, correlating with clinical resolution of pain.

HISTOLOGY

Thirty six patients underwent excision biopsy of

their lesions, and histological confirmation of the diagnosis of osteoid osteoma was obtained. Microscopically all biopsies presented a characteristic picture with a well circumscribed nidus of osteoid and woven bone surrounded by regular plump fibroblasts, osteoblasts, and occasional osteoclastic giant cells. Between the trabeculae, there was a background stroma of vascular fibrous tissue. Two patients who presented as neurological problems (cases 1 and 2) had muscle biopsies carried out to investigate their muscle wasting. In case 1 a needle biopsy specimen from vastus lateralis showed predominance of type 1 fibres with hypertrophy of type 2A and 2B fibres, consistent with a 'fibre type disproportion'. Case 2 had an open muscle biopsy specimen taken from adductor longus, which showed only mild atrophy of type 1 and 2A fibres. Although probably within normal limits for this muscle, a neuropathic process could not be entirely excluded.

Discussion

An osteoid osteoma is a benign neoplasm of bone with distinctive clinical signs and symptoms. It is between two and three times as common in boys as in girls, with a maximum age incidence in the second and third decades of life.^{1 2} The bones most commonly affected are the femur and tibia, the lesions usually being located near the shaft. In the vertebrae, the most common site is the neural arch.² Almost without exception the initial symptom is pain, which is vague and intermittent at first but gradually increasing in severity. The pain is described as boring or aching in character, and is characteristically worse at night and relieved by salicylates.¹

Despite these classical features, the clinical diagnosis of osteoid osteoma can be notoriously difficult. The interval between the onset of symptoms and a correct diagnosis being made is seldom less than six months and it may be many years.¹ In our series the mean duration from the onset of symptoms to diagnosis was 13.8 months. Delayed diagnosis may be explained by the presence of atypical clinical features, particularly neurological, and from lack of aware-

ness that plain radiographs in the early stages can be normal. Although the characteristic pain is usually felt in the immediate region of the bone lesion, both 'referred' and 'radicular-like' pain are well described.³⁻⁶ Referred pain arises commonly from three sites: (i) the lumbar spine, especially when the pars interarticularis is affected—pain is referred to the abdomen or into the leg; (ii) the region of the lesser trochanter of the femur—pain is commonly referred to the knee⁸; and (iii) the posterior aspect of the upper third of the tibia—pain is referred up the thigh to the hip.⁹ In our series, three patients with femoral osteomas experienced only referred pain in the knee without local pain, thereby misdirecting radiological studies. One patient (case 36) with an osteoma in the left neural arch of S2 complained of buttock and thigh pain. Rare cases of painless, histologically diagnosed, osteoid osteomas have also been reported.¹⁰⁻¹²

The fact that osteoid osteomas may mimic certain neurological diseases, in particular spinal lesions, may also be a cause of delayed diagnosis resulting in unnecessary neuroradiological studies. Localised atrophy of some muscles of the affected limb, decreased deep tendon reflexes, weakness, and rarely dermatomal sensory loss, are all well described.^{1 3-5} Rushton *et al* also found abnormal straight leg raising in two patients with lesions of the femoral shaft.³ Halperin *et al* reported two children with osteoid osteomas of the proximal femur simulating spinal root compression,⁴ and Young included four patients with femoral osteomas in his review of non-neurological lesions simulating protruded intervertebral disc.⁵

Radiologically an osteoid osteoma can usually be seen as a well defined area of dense sclerosis with a diameter of 0.5–1 cm in the centre of which is a lucent nidus, which may contain speckled areas of calcification.^{1 13} Symptoms can precede the development of any radiological evidence of the tumour. Cortical osteoid osteomas usually result in cortical thickening and a zone of sclerosis, but subperiosteal lesions and those in cancellous bone produce much less sclerosis and this may not be evident radiologically.^{2 13} Six of our patients had normal plain radiographs of the relevant regions, five of whom had femoral neck osteomas. Lesions in this region, by virtue of the intracapsular location and cancellous nature of the bone, produce little sclerosis.² This, in addition to the normal irregularities in density and margins of the proximal end of the femur, can result in diagnostic difficulty. Radionucleotide scanning is indicated if the possibility of the diagnosis is borne in mind and if the plain radiographs are normal. If plain films are abnormal but not diagnostic, a computed tomogram with fine slices through the relevant region is recommended (fig 2).

The histological appearance of an osteoid osteoma varies with the age of the lesion and with its site.⁷ The characteristic pain is presumed to be related to the extremely vascular nature of the lesion, and increased tension and oedema within the lesion are thought to cause the pain by direct stimulation of local nerves around the intraosseous vessels. Sherman and

Figure 2 Case 12: computed tomogram showing cortical thickening of the left femoral neck medially, a lucent area within the cortex, and a calcified nidus (arrow).

McFarland showed that there were unmyelinated nerve fibres in the fibrous zone around the nidus, and implicated them as the mediators of the pain.¹⁴ Schulman and Dorfman, using a specific silver stain, showed that there were nerve fibres within the nidus itself.¹⁵ These fibres accompanied blood vessels and were presumed to be derived from the autonomic nervous system.¹⁴ The spinal origin of such fibres implies they can conduct pain in a radicular distribution, which accounts for the reported 'radicular syndromes' associated with these lesions,⁴ and the occasional local vasomotor phenomena characterised by increased temperature and hyperhydrosis.⁸ Makley and Dunn found increased prostaglandin synthesis, particularly of prostaglandin E₂, in three osteoid osteomas when compared with normal bone, and postulated that this may cause intense local pain by vasodilatation and hyperalgesia.¹⁶

Treatment of osteoid osteoma consists of en bloc excision of the tumour and some surrounding bone. Complete removal of the nidus is invariably accompanied by almost immediate relief of pain.^{1 2} Two patients (cases 26 and 36) required reoperation for recurrent pain; in one of them initial histopathological examination failed to confirm excision of the tumour nidus. One patient (case 6) who had a femoral neck osteoma, had the lesion successfully excised by a percutaneous technique under computed tomographic guidance. Prognosis from surgical excision was equally favourable in those patients presenting with neurological symptoms and signs as in those presenting with pain alone. The natural history of osteoid osteomas is difficult to document because without biopsy the diagnosis cannot be proved, and biopsy may change its normal development. Indirect evidence suggests, however, that the condition runs a self limiting course.^{7 17 18} In our series, case 14 did not undergo surgical excision of the lesion. Follow up imaging studies 15 months after diagnosis showed obvious healing of the lesion, correlating with clinical resolution of the pain.

Diagnosis of osteoid osteoma requires an accurate clinical history and an awareness of the possibility of the diagnosis. The simultaneous muscle atrophy and decreased deep tendon reflexes in the affected limb may erroneously suggest the presence of a primary neurological disorder. Awareness of this, and of the occurrence of radicular and referred pain, may avoid unnecessary neuroradiological investigations. In the context of a highly suggestive clinical history, radionucleotide scanning should be undertaken even if plain radiographs are entirely normal.

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