

## Osteoid osteoma in a 16-year-old boy presenting with atrophy of the left thigh: diagnostic difficulties

Sinem Akgül<sup>1</sup>, Akın Üzümcügil<sup>2</sup>, M. Fani Bozkurt<sup>3</sup>, Meral Topçu<sup>1</sup>

Departments of <sup>1</sup>Pediatrics, <sup>2</sup>Orthopedics and Traumatology, and <sup>3</sup>Nuclear Medicine, Hacettepe University Faculty of Medicine, Ankara, Turkey

**SUMMARY:** Akgül S, Üzümcügil A, Bozkurt MF, Topçu M. Osteoid osteoma in a 16-year-old boy presenting with atrophy of the left thigh: diagnostic difficulties. Turk J Pediatr 2008; 50: 373-376.

Osteoid osteoma is an osteoblastic benign lesion of the bone. The pathognomonic symptom is significant pain, which responds well to nonsteroidal anti-inflammatory drugs. When typical clinical and radiological features are present, the diagnosis is not difficult. However, if the lesion is in an area not clearly seen on plain radiographs, or clinical features are atypical, then diagnosis becomes difficult. We present a case of osteoid osteoma with delayed diagnosis that presented itself with neurological signs. Prominent features present in the patient included pain that responded well to medication and muscle atrophy, which led to a wider differential diagnosis. Diagnosis was made approximately two years after the onset of his initial symptoms, after having been investigated and treated both in our own hospital and elsewhere. This case illustrates clinical and radiological diagnostic problems of osteoid osteoma, demonstrating that it can present itself with neurological signs. Correct diagnosis then requires detailed history and clinical awareness.

**Key words:** osteoid osteoma, limb atrophy, neurological signs, delayed diagnosis.

Osteoid osteoma was first described by Bergstrand in 1930 and was characterized by Jaffe as an entity in 1935<sup>1</sup>. It is an osteoblastic benign lesion of the bone, usually occurring in a long bone cortex, leading to marked cortical thickening. It is commonly seen between 10-30 years of age with a male to female ratio of 2-3:1<sup>1</sup>. It characteristically has a radiolucent nidus on radiographs that is believed to be responsible for severe pain, as evidenced by the disappearance of pain when it is completely excised<sup>2</sup>. The lower limb long bones are the most frequent sites; however, osteoid osteomas have been reported in many other sites, such as the pelvis, foot and hand bones, and the vertebra, and at more than one site (metachronous or multicentric)<sup>3,4</sup>. In most cases, affected individuals complain of severe pain increasing at night and pain relief with use of nonsteroidal anti-inflammatory drugs (NSAIDs)<sup>5</sup>. Diagnosis of osteoid osteoma is not difficult when the typical clinical and radiological features are present. However, if the lesion is in an area not clearly seen on plain radiographs, or the clinical features are atypical, the diagnosis becomes more difficult<sup>6</sup>.

We present a patient with osteoid osteoma who was diagnosed two years after the start of his pain, having previously been treated and investigated both in our own hospital and elsewhere. The only other positive finding was the atrophy of the involved limb, which led to a wider differential diagnosis. The aim of this case report is to illustrate some of the clinical and radiological diagnostic problems of osteoid osteoma and to increase awareness of its less common clinical findings delaying diagnosis.

### Case Report

A 16-year-old boy presented with a history of left thigh pain that radiated to the lateral aspect of the thigh and left knee. His pain had started two years ago, but over time the pain had increased in intensity. He stated that his pain was so bad that he had stopped going to school, spent most of his time in bed, and was awakened nearly nightly because of the pain; hence, various NSAIDs were prescribed. He reported that these drugs had provided significant pain relief and that after

taking the medication, his pain disappeared in approximately 15 minutes. The patient's father had a history of multiple sclerosis.

On physical examination, we found normal ranges of motion of lumbar vertebrae and of hip and knee joints, and the neurological examination was also normal. The only pathological finding was the atrophy of his left thigh. A 5 cm difference was determined between thigh measurements (right: 44.5 cm, left: 39.5 cm) (Fig. 1a, b).

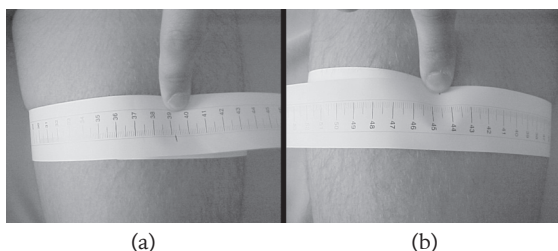


Fig. 1a-b. Photographs demonstrating atrophy of the left thigh.

The anteroposterior, lateral spinal and antero-lateral pelvic radiographs showed no sign of radiolucency or osteosclerosis (Fig. 2). His cranial and spinal magnetic resonance imagings were found to be normal, as was his left leg electromyogram. Because of the difference in thigh circumferences, an abdominal ultrasonography was performed for the exclusion of a possible malignancy that can be seen with overgrowth syndromes<sup>7</sup>, and it was found to be normal. Bone marrow aspiration material was obtained<sup>8</sup> and also found to be normal. Diagnostic work-up, as a complete blood count and acute phase reactants, for a possible bone infection, yielded negative results. The next investigation was a three-phase bone scintigraphy with Tc99m-MDP (methylene diphosphonate) that showed an increased uptake of radioactivity

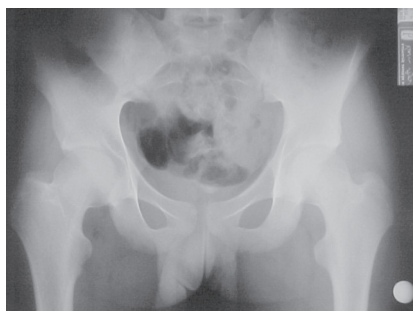


Fig. 2. The normal pelvic radiograph.

at the collum (neck) of the femur, which was consistent with a benign or malignant bone lesion, a bone infection or fracture (Fig. 3). A subsequent pelvic computerized tomography (CT) showed the presence of a nidus at the superior intertrochanteric part of the neck of the femur (Fig. 4). The patient was consulted by an orthopedic surgeon and CT- guided radioablation of the lesion was planned.



Fig. 3. The bone scan showing an increased uptake of the isotope at the neck of the left femur.



Fig. 4. Pelvic CT showing the presence of the osteoid osteoma at the superior intertrochanteric part of the neck of the femur.

## Discussion

Classically, patients with osteoid osteoma present with a history of dull pain in a long bone lasting for several months<sup>9</sup>. Prominent features present in patients included pain that responds well to medication. The interval between the onset of symptoms and eventual diagnosis is commonly 6 to 12 months and sometimes even years. In our case, diagnosis was made approximately two years after the onset of his initial symptoms. The diagnosis of osteoid osteoma is often difficult in children and the reasons for the delay in diagnosis include mainly nonspecific and misleading neurological symptoms and absence of findings on radiologic examination<sup>10</sup>.

Patients with osteoid osteomas of the femur or tibia may present with neurological manifestations mimicking spinal cord lesions, radiculopathies, and peripheral nerve disorders. Diagnostic testing is often aimed at excluding these possible neurological etiologies, causing a delay in the diagnosis and leading to muscle atrophies, diminished deep tendon reflexes, tenderness, localized swelling or contractures in the process<sup>6,10,11</sup>. The presence of a focal neurological sign in our patient was misleading and caused a delay in the diagnosis. In the study by Kiers et al.<sup>12</sup> describing the clinical and radiological features of 38 children with osteoid osteomas, 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 localized muscle atrophy, and 10 patients (34%) had diminished or absent deep tendon reflexes in the affected limb. Six patients had normal plain radiographs.

In our case, the hemiatrophy brought about the question of whether this was a true atrophy of the left thigh or a hemihypertrophy of the right. Hemihypertrophy is commonly related to malignancies like Wilms' tumor or hepatoblastoma, which can also cause pain because of the pressure of the tumor<sup>7,13</sup>, and is why the abdominal and pelvic ultrasonography were performed. Since hematological malignancies<sup>9</sup> are another common cause of bone pain in children, the bone marrow material was evaluated. Both of these decisions delayed the diagnosis, leading to extra diagnostic testing in seeking the etiology. We believe that atrophy was caused by the consequent disuse of the affected limb because of pain. Skeletal muscle atrophy may occur following surgery, immobilization, non-weight-bearing, and extended periods of bed rest. We learned that after the pain began, our patient stopped going to school and spent most of his time in bed. Disuse muscle atrophy is debilitating and increases the risk of future health problems such as hip fractures, cardiovascular deconditioning, metabolic derangement, osteoporosis, and decreased functional independence<sup>14</sup>.

The complex three-dimensional anatomy of the pelvis leads to difficulty in visualization of the lesion. For the body regions such as spine,

pelvis and proximal femur, it is difficult to reveal the pathognomonic nidus formation with conventional plain radiographs<sup>2</sup>. On the plain radiographs taken in both our clinic and the clinic before, the nidus could not be visualized. In patients suspected of an osteoid osteoma, radionuclide bone scintigraphy is thought to be a more sensitive method<sup>9</sup>. In such cases, bone scans commonly show a double-density sign with increased uptake centrally that distinguishes the lesion from osteomyelitis or abscess<sup>2,15</sup>. The positive bone scan in our case proved to be helpful in the diagnosis, but since the increased uptake of radionuclide material was diffuse and nonspecific, there was need for further investigations. Considering the efficiency of CT in the demonstration of internally located sclerotic nidus, we performed a CT of the left hip and succeeded in demonstrating the osteolytic region with a sclerotic nidus located inside<sup>16</sup>.

There are three main approaches for the treatment of osteoid osteoma, namely: surgical excision, conservative (medical) treatment, and percutaneous excision. Osteoid osteoma has been traditionally treated with conservative methods (NSAID), surgical excision or curettage, but newly developed minimally invasive techniques, such as CT-guided percutaneous radiofrequency thermal ablation and laser photocoagulation, have become the methods of choice for the treatment<sup>17,18</sup>. In the present case, CT-guided radioablation of the nidus was planned.

Whether or not the atrophy will disappear after treatment is one of the frequently asked questions before the initiation of therapy. A study by Hsich et al.<sup>10</sup> showed that one month after heat ablation treatment of the osteoid osteoma, the muscle atrophy had resolved, and the circumference of the thighs were symmetric in their patient. Deep tendon reflexes were also equal, although the leg length discrepancy was still present. Interestingly, nine months after the ablation treatment, this leg length discrepancy had resolved as well.

In summary, pain is the most common symptom of osteoid osteoma and is characteristically quite responsive to even mild analgesics; however, focal neurological signs, including weakness, atrophy, and diminished deep tendon reflexes, make diagnosis difficult. CT is likely

the most accurate imaging modality. In order to minimize delay in diagnosis in osteoid osteoma, we conclude that it may be prevented with the knowledge that concomitant muscle atrophy and depressed deep tendon reflexes are relatively common findings, and that the characteristic radiological features may only appear late in the disease course. Most importantly, proper diagnosis requires a clinical awareness of the unusual issues involved with this entity.

#### REFERENCES

1. Lee EH, Shafi M, Hui JH. Osteoid osteoma: a current review. *J Pediatr Orthop* 2006; 26: 695-700.
2. Duman I, Aydemir K, Tan AK, Dinçer K, Kalyon TA. An unusual case of osteoid osteoma clinically mimicking sacroiliitis. *Clin Rheumatol* 2006 (Epub) DOI 10.1007/s10067-006-0280-8.
3. Radcliffe SN, Walsh HJ, Carty H. Osteoid osteoma: the difficult diagnosis. *Eur J Radiol* 1998; 28: 67-79.
4. Sinha S, Housden P. Discrete synchronous multifocal osteoid osteoma of the femur. *Pediatr Radiol* 2004; 34: 280.
5. Mungo DV, Zhang X, O'Keefe RJ, Rosier RN, Puzas JE, Schwarz EM. COX-1 and COX-2 expression in osteoid osteomas. *J Orthop Res* 2002; 20: 159-162.
6. Franceschi F, Marinozzi A, Papalia R, Longo UG, Gualdi G, Denaro E. Intra- and juxta-articular osteoid osteoma: a diagnostic challenge. Misdiagnosis and successful treatment: a report of four cases. *Arch Orthop Trauma Surg* 2006; 126: 660-667.
7. Lapunzina P. Risk of tumorigenesis in overgrowth syndromes: a comprehensive review. *Am J Med Genet C Semin Med Genet* 2005; 137C: 53-71.
8. Jones OY, Spencer CH, Bowyer SL, Dent PB, Gottlieb BS, Rabinovich CE. A multicenter case-control study on predictive factors distinguishing childhood leukemia from juvenile rheumatoid arthritis. *Pediatrics* 2006; 117: 840-844.
9. Frassica FJ, Waltrip RL, Sponseller PD, Ma LD, McCarthy EF. Clinicopathologic features and treatment of osteoid osteoma and osteoblastoma in children and adolescents. *Orthop Clin North Am* 1996; 27: 559-574.
10. Hsich GE, Davis RG, Darras BT. Osteoid osteoma presenting with focal neurologic signs. *Pediatr Neurol* 2002; 26: 148-152.
11. Alani WO, Bartal E. Osteoid osteoma of the femoral neck simulating an inflammatory synovitis. *Clin Orthop Rel Res* 1987; 223: 308-312.
12. Kiers L, Shield LK, Cole WG. Neurological manifestations of osteoid osteoma. *Arch Dis Child* 1990; 65: 851-855.
13. Finch GD, Dawe CJ. Hemiatrophy. *J Ped Orthop* 2003; 23: 99-101.
14. Frimel TN, Kapadia F, Gaidosh GS, Li Y, Walter GA, Vandenborne K. A model of muscle atrophy using cast immobilization in mice. *Muscle Nerve* 2005; 32: 672-674.
15. Schaefer MP, Smith J. The diagnostic and therapeutic challenge of femoral head osteoid osteoma presenting as thigh pain: a case report. *Arch Phys Med Rehabil* 2003; 84: 904-905.
16. Assoun J, Richardi G, Railhac JJ, et al. Osteoid osteoma: MR imaging versus CT. *Radiology* 1994; 191: 217-223.
17. Ghanem I. The management of osteoid osteoma: updates and controversies. *Curr Opin Pediatr* 2006; 18: 36-41.
18. Rosenthal D, Hornicek F, Torriani M, Gebhardt MC, Mankin HJ. Osteoid osteoma: percutaneous treatment with radiofrequency energy. *Radiology* 2003; 229: 171-175.