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Solid Tumour Section

Mini Review

Bone: Osteoid Osteoma

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Identity

Note

Osteoid osteoma (OO) is defined as a benign bone-forming tumour characterized by small size, limited growth potential and disproportionate pain (WHO, 2002). Usually, OO is solitary, although the literature shows few reports of multiples lesions in the same patient, but not concomitant.

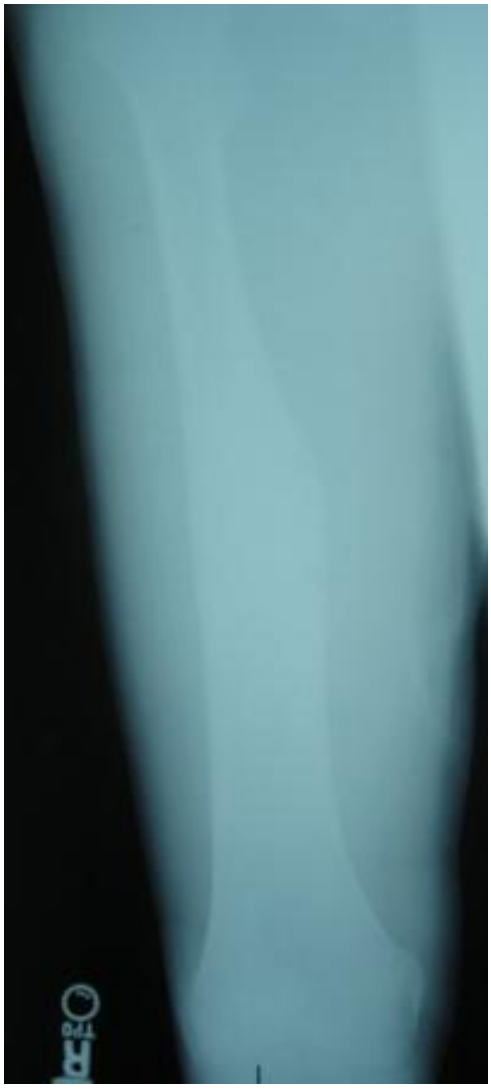
In most cases, OO patients complain of severe nocturnal pain that usually responds to salicylates and nonsteroidal anti-inflammatory drugs (NSAIDs). The exact mechanism of the pain in OO and why it is more intense at night is still unknown. Some reports have shown that the nidus is responsible for the severe pain, as evidenced by the disappearance of pain when the nidus is completely excised, and that prostaglandins seem to play a major role. Prostaglandin E2 is believed to prompt vasodilation and pain. An increase concentration of prostaglandin has been found in the nidus of the lesion, as well as, a strong positivity for cyclooxy-genase-2 (a key enzyme in the production of prostaglandins, particularly prostaglandin E2).

Clinics and pathology

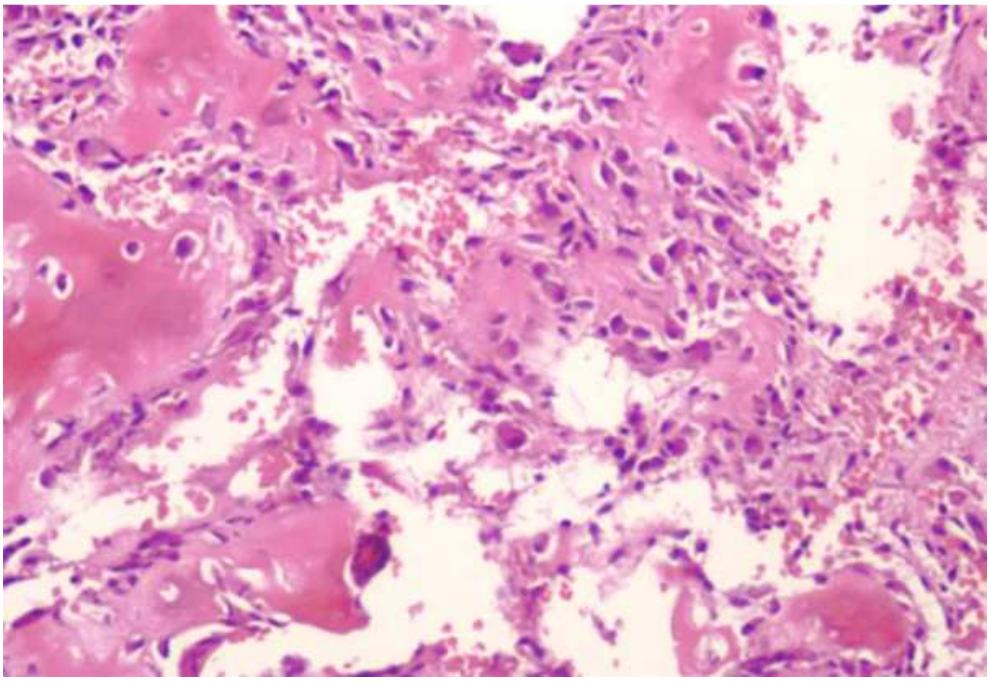
Etiology

The pathogenesis of OO remains controversial. Some hypotheses have been reported:

OO is a real neoplasm; OO is a reactive process that arises on an inflammatory basis, OO is an unusual healing and reparative process. However, this tumour does not enlarge, seldom exceeds 1cm in greatest diameter; and in some cases, spontaneous regression and healing occur. Contrary to typical neoplasms, the nidus of OO contains a relatively immature central area with more mature calcified peripheral bone.



Osteoid Osteoma. Showing dense surrounding reactive sclerotic bone.



Osteoid Osteoma is characterized by anastomosing woven bone trabeculae, rimmed by a single layer of prominent osteoblast.

Epidemiology

OO is the third most common benign bone tumor. Its incidence is 11% among the benign tumors and 3% among all primary bone tumors. It is most common in the second and third decades. There is a male to female preponderance of 3 to 1.

The majority of the OO cases occur in lower extremity of long bones, particularly in the proximal femur. In the spine, they almost exclusively occur in the posterior vertebral elements. Additionally, OOs may occur in a juxtaarticular bone within a synovial cavity where they are termed intraarticular OOs. They are typically located in diaphyseal or metaphyseal cortices, but may rarely be located in the medullary cavity or periosteum. Infrequently, they are seen in the epiphysis.

Clinics

Characteristically, patients complain of localized pain, generally worse at night and relieved with salicylates. This classic presentation is not always present and, however, night pain may be observed with other bone tumours. More importantly, virtually all patients have progressive pain.

The natural history of untreated OO is toward spontaneous regression, it is taken an average of 6 years. During this period, the nidus gradually begins to calcify; afterwards, ossify, and, finally, blends into sclerotic surrounding bone. The local pain gradually diminishes.

Indication for surgery depends on symptom severity and the patient's pain tolerance. With surgical resection, it has been emphasized that nidus removal is essential for pain relief.

Familial occurrence of OO is an exceedingly rare event. The literature has reported only two descriptions of OO occurring in two siblings (Kaye and Arnold, 1977; Kalil and Antunes, 2003).

Pathology

Morphologically, OO is a well-circumscribed lesion, consisting of a central area of woven bone trabeculae and osteoid, lined by a prominent single layer of plump osteoblast, and an intervening hypocellular but highly vascular stroma. The bone trabeculae are irregularly arranged and thin. Hyaline cartilage is not present. Osteoclasts may be distributed randomly throughout the lesion. Significant mitotic activity is usually absent and, when preset, limited to the stromal cells. Mature OO may be composed virtually entirely of compact woven bone. In all cases, the nidus is sharply demarcated from the surrounding, generally sclerotic bone.

The nidus of OO usually has 1.5 to 2 cm in size and consists of osteoid, osteoblasts, and variable amounts of fibrovascular stroma.

O'Connell et al., in 1998, reported that 25 of 34 cases of OO contained phosphorylated neuro-filament, -neurofilament-, and/or S-100-positive nerve fibers in the reactive zone around the nidus and/or in the nidus. The nerve fibers were larger and more abundant in the reactive zone than in the nidus, and they were occasionally visible on HE-stained slides, on retrospective review. None of the control "bone tumours" (e.g., osteoblastomas, osteosarcomas, giant cell tumours) had detectable nerve fibers either within their substance or within the surrounding bone.

Treatment

After radiographic and surgical localization, the nidus should be removed in its entirety.

Prognosis

Local recurrence is rare but may occur years after the initial surgery.

Genetics

Note

Baruffi et al., in 2001, described the cytogenetic analysis of cells derived from two OO. The first tumour showed del(22)(q13.1) as the sole clonal chromosome alteration. The same alteration was present in the second OO, as a non-clonal entity. The authors mentioned that structural chromosomal alterations involving 22q13.1 in OO may affect critical genes involved in the regulation of cell proliferation, such as the YWHAH gene that codes for a 14-3-3 family members, dimeric phospho-serine-binding proteins that participate in signal transduction and checkpoint control pathways. Their primary function in mammals is to inhibit apoptosis. They also described that another gene mapped in this region is PDGFB that codes for a platelet-derived growth factor, a beta polypeptide (simian sarcoma viral [v-sis] oncogene homolog), a potent mitogen for cells of mesenchymal origin, also involved in the transformation process.

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