Pearls & Oy-sters:
Osteoid osteoma of the scapula masquerading as neuralgic amyotrophy

PEARLS

• Osteoid osteoma is a benign bone tumor that usually presents in children and young adults characterized by local pain which is worse at night; occasionally the pain is referred to a site distant from the actual lesion.
• Osteoid osteoma of the scapula should be considered in the differential diagnosis of neuralgic amyotrophy due to shared clinical features of pain, weakness, and atrophy around the shoulder and upper arm.
• Osteoid osteoma is often amenable to cure by minimally invasive procedures particularly in locations like the scapula and pelvis where open surgical procedures could lead to significant complications.

OY-STER

• The diagnosis of osteoid osteoma may be delayed by months to years in children due to young age, focal neurologic symptoms and signs, and absence of characteristic findings on radiologic examination at an early stage.

CASE REPORT

A 7-year-old previously healthy right-handed boy presented to the pediatric neurology clinic with right arm and shoulder pain of 4 months duration. The pain was severe in nature, could last for an hour, at times woke him up from sleep, and was partially relieved with acetaminophen and ibuprofen. About 4 weeks prior to presentation, his parents noticed that his right upper arm was thinner than the left. The patient denied any sensory loss or weakness of the right arm or other limbs. There was no history of recent trauma or immunizations. His birth and developmental history were unremarkable and there was no family history of neuromuscular disorders. On clinical examination, his higher mental function and cranial nerves were normal. Motor system examination revealed atrophy of the right upper arm. There was no shoulder drooping, scapular winging, wasting of the forearm and hand, or fasciculations of the muscles. There was no local tenderness, redness, or swelling and range of motion of the shoulder joint was normal. There was only mild weakness of the muscles around the right shoulder joint (4+/5). The right biceps, triceps, and brachioradialis tendon reflexes were depressed compared to the left. The sensation was normal in the right upper limb. The initial clinical impression was that of neuralgic amyotrophy. Extensive EMG study of the right arm with additional nerve conduction studies (NCS) showed no definite evidence of a right brachial plexopathy, shoulder girdle neuropathy, radiculopathy, or motor axon loss process. The normal electrodiagnostic studies triggered a search for other conditions which could mimic neuralgic amyotrophy:1 cervical radiculopathy, mononeuritis multiplex, multifocal motor neuropathy, and brachial amyotrophic diplegia including Hirayama disease (as rarely neuralgic amyotrophy could be painless, manifesting with motor features only). However, all these processes are quite rare or not described in children of this age and furthermore the NCS and EMG ruled out all these entities. MRI of the cervical spine ruled out spinal cord pathology. The non-neurologic conditions which could masquerade as neuralgic amyotrophy include shoulder joint pathology and complex regional pain syndrome. The normal range of motion rules out shoulder joint pathology. Complex regional pain syndrome is unlikely given the lack of allodynia, and the lack of any skin temperature or color differences. X-ray of the shoulder and humerus was normal. As the pain was persistent further imaging studies including CT scan of the chest was performed to look for any ribcage or bony pathology that might lead to referred pain. The CT scan revealed a tiny lucency in the right scapula, suspicious for an osteoid osteoma (figure 1). A subsequent MRI showed a 9-mm osteoid osteoma in the scapular body which was in close proximity to the brachial plexus (figures 2 and 3) raising the concern for potential brachial plexus injury. He underwent CT-guided radiofrequency ablation of the lesion.

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guided biopsy performed from the lesion ruled out osteoblastoma (a benign bone tumor which can mimic osteoid osteoma) or malignancy. He tolerated the procedure well with complete disappearance of the painful neuralgia within weeks. At 4 months follow-up there was almost complete resolution of the wasting of the right upper arm with normal strength and retained reflexes.

**DISCUSSION**

Osteoid osteoma is a benign osteogenic tumor comprising 10%–12% of all benign bone tumors occurring predominantly in children and young adults and is commoner in males. The majority of the lesions affect the long bones (femur, tibia). Involvement of the flat bones is relatively uncommon. Osteoid osteomas of the scapula are very rare. Osteoid osteoma is typically a small lesion (<1.5 cm in diameter) and consists of a central nidus, composed of variably calcified bony trabeculae on a background of fibrous, vascular, and nerve tissue.

The most common presenting symptom is local pain, typically more severe at night, and classically responds to salicylates. Although the characteristic pain is usually felt in the immediate vicinity of the lesion, both referred and radicular-type pain are well-described. The stimulation of the unmyelinated nerve endings in the nidus by the marked vascularity may be the cause for pain. A marked local increase in prostaglandin synthesis suggests that the pain is caused by vasodilation in the nidus.

Osteoid osteoma may mimic neurologic diseases due to localized atrophy of muscles of the affected limb, focal weakness, and decreased deep tendon reflexes. Muscle weakness and atrophy may be caused by disuse atrophy due to severe pain. The depressed reflexes are possibly due to focal weakness and muscle atrophy, although this finding is less clearly understood. The diagnosis of osteoid osteoma is often difficult in young children as there is a considerable lag between the symptom onset and the eventual diagnosis.

The radiologic hallmark of osteoid osteoma on plain X-ray is usually an oval radiolucency representing the nidus as well as a surrounding area of reactive bone sclerosis with or without periosteal new bone formation. Symptoms can precede the radiologic evidence of the tumor, so it can be missed at an early stage. Radionucleotide scanning showing increased uptake can be considered if the suspicion is strong. CT scan with thin slices through the relevant region is usually diagnostic. In our patient CT of the chest raised the suspicion of osteoid osteoma which was confirmed on MRI scan.

Neuralgic amyotrophy previously known as Parsonage-Turner syndrome is a distinct peripheral nervous system disorder characterized by episodes of extreme pain at onset, rapid multifocal paresis and atrophy of the upper extremity muscles, and a slow recovery requiring months to years. Patchy areas of hypesthesia are usually present. The upper trunk of the brachial plexus is most commonly involved, but any part can be affected. The presence of severe pain at onset with atrophy and mild weakness in the distribution of the upper trunk of the brachial plexus led us to a diagnosis of neuralgic amyotrophy in our
The pain in neuralgic amyotrophy is often worse at night, a feature shared by osteoid osteoma. The initial pain usually lasts about 4 weeks, though in 10% of individuals it can last beyond 2 months. In our patient the pain was persistent beyond 4 months, an atypical feature for neuralgic amyotrophy. Furthermore electrodiagnostic studies are always abnormal in neuralgic amyotrophy in contradistinction to our patient. The true diagnosis was delayed in our patient as the pain was referred to the shoulder region and upper arm coupled with focal neurologic findings pointing to brachial plexopathy, radiculopathy, or a spinal cord lesion. Osteoid osteoma of the scapula should be considered in the differential diagnosis of neuralgic amyotrophy particularly in children and young adults.

Successful therapy of osteoid osteoma constitutes total removal or destruction of the nidus. Intraoperative localization of these small lesions can at times be very challenging and open surgical removal of the tumor often requires significant bone destruction and subsequent bone grafting. Minimally invasive therapies designed for osteoid osteoma aim to achieve removal or destruction of the nidus with minimal tissue invasion. CT-guided radiofrequency thermal ablation has been proven to be an accepted, safe, minimally invasive, and cost-effective treatment for osteoid osteoma. Percutaneous CT-guided procedures may reduce morbidity and complications when compared with traditional open surgical resection, especially in unusual locations such as the scapula.

Confirming the diagnosis of osteoid osteoma is important as these lesions are curable with minimally invasive therapy with complete resolution of all neurologic deficits.

AUTHOR CONTRIBUTIONS
P. Ghosh collected and organized the data and wrote the first manuscript (including the first draft). M. Moodley conceptualized the study. M. Moodley and S. Mitra verified the results and revised the manuscript at all stages.

DISCLOSURE
The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES
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