A case report of myositis ossificans progressiva complicated by femoral nerve compression treated with radiotherapy

Sir, We present a female who was noted to have abnormal toes from birth. At age 19 yr she developed a lump below her jaw and lumps in both shoulders and the left breast with progressive musculoskeletal stiffness and pain. A biopsy of one lesion revealed fibrosis, focal muscle destruction and inflammation. Myositis ossificans progressiva was then diagnosed.

She presented aged 34 yr with a 5-week history of right thigh pain, fixed flexion deformity of the thigh causing inability to extend the knee, and numbness of the right leg. The right knee jerk was absent and there was sensory disturbance in the territory of the saphenous branch of the femoral nerve. Her erythrocyte sedimentation rate (ESR) was 22 mm/h. Ultrasonography showed a hypoechoic mass posterolateral to the right femoral neurovascular bundle. Surface EMG demonstrated a femoral neurapraxia. CT scanning (Fig. 1) showed swelling of the iliopsoas muscle, possibly involving the femoral vessels and nerve. She received physiotherapy, indomethacin (150 mg daily), disodium etidronate (400 mg/kg daily) and a single fraction of radiotherapy (dose 10 Gy) to the abnormal area seen on the scan. Two months later, her pain had completely subsided, as had the signs of neurapraxia. A repeat CT scan demonstrated resolution of the muscle oedema and swelling, but the presence of calcification.

Myositis ossificans progressiva (also known as fibro-dysplasia ossificans progressiva) is a severe, rare condition of ectopic ossification with primary involvement of the skeletal muscles, associated with characteristic...
skeletal abnormalities [1]. It has autosomal dominant inheritance with complete penetrance but variable expressivity, and most cases result from a sporadic mutation [2]. Genes for bone morphogenetic proteins, in particular BMP4, are thought to be plausible candidate genes [3].

Ectopic ossification usually starts in early childhood. Radiological evidence of ossification is not usually present until 4–6 weeks after the appearance of a lump. Sites of ossification include the neck, spine and shoulder girdle. Trauma precipitates new lesions. The ESR may be elevated during an acute episode. Radiological abnormalities of the toes and thumbs are common.

Intraneural ossification is uncommon. Nerve entrapment is unreported and has been described in cases of heterotopic ossification due to other causes [4].

In early disease, radiographs may be normal or show soft-tissue swelling. Weeks later, films show ectopic calcification. Ultrasonography of early lesions may show a sonolucent soft tissue mass with echogenic surrounding oedema; calcification within the lesion may also be seen. CT is sensitive for calcification, while MRI of early lesions shows high signal intensity on T2-weighted images associated with proliferating fibroblasts. Early calcification is not typically detectable. Late MRI scans show low T1 and T2 signals associated with fibrous or calcified tissue.

Histologically, early lesions resemble granulation tissue, occasionally with cellular inflammatory infiltrate. Spicules of bone appear in the centre of the fibroblastic nodules. Bone and cartilage formation is seen in mature specimens. The bone formed initially is of the woven type; this is later remodelled to mature lamellar bone.

There is little convincing evidence that any form of treatment alters the progress of the disease in myositis ossificans progressiva. Treatments that have been used include a diet reduced in vitamin D and calcium, the avoidance of sunshine, and treatment with corticosteroids. Other treatment strategies include beryllium, vitamins B and E and penicillamine [1]. Drugs that block calcification have also been used, including EDTA (disodium ethylene diamine tetraacetic acid), isotretinoin [1] and etidronate [5], without proven benefit. There is one report of treatment with ascorbic acid [6]. Surgical resection of the ossified sites is usually unsuccessful, with recurrence at the same site, but occasionally good outcomes have been described after surgery [7].

Management strategies used for ossification after hip surgery and spinal cord injury may be useful. Various treatment modalities have been described, including the use of NSAIDs (non-steroidal anti-inflammatory drugs) (particularly ketorolac and indomethacin), warfarin, radiotherapy and surgical resection [8]. Etidronate has been used to treat heterotopic ossification after spinal cord injury [9] and its use in myositis ossificans progressiva has also been documented in case reports [5]. Low-dose radiotherapy has not been described in myositis ossificans progressiva, but has in patients following spinal injuries, for the prophylaxis of ossification at the time of hip surgery, and to treat the postoperative formation of new bone. This treatment has appeared effective without side-effects [10].

We have described the case of a 34-yr-old woman with neurapraxia secondary to myositis ossificans progressiva. This disease has no adequate treatment, though low-dose radiotherapy in combination with other treatment strategies may reduce entrapment pain and increase mobility without significant side-effects.

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