Case Reports and Technical Notes

Dystrophic Muscle Calcification Following Cerebral Damage

Report of Three Cases

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DYSTROPHIC muscle calcification in patients with paraplegia is well recognized, but has rarely been observed in association with intracranial lesions. Voss made a comprehensive classification of muscle calcification in neurological disorders and described three cases associated with intracranial lesions. Seven subsequent cases have been reported. The three additional patients described here had multifocal lesions, and in one instance, the abnormal calcification almost completely resolved.

Case Reports

Case 1. A 44-year-old Nigerian man was admitted to the hospital in January, 1962, with a severe headache. He subsequently became comatose, and investigations showed an aneurysm at the beginning of the right middle cerebral artery. Following a craniotomy and ligation of the neck of the aneurysm, he developed a left hemiplegia and did not fully regain consciousness for 2 months.

In April while still in the hospital his knees became stiff, and a bony-hard mass was noted in the right thigh. X-ray films showed "myositis ossificans" around both hips, around the right femoral shaft, and left knee (Fig. 1). The serum calcium was normal (10.0 mg/100 ml) as was the urinary calcium (116 mg daily). The patient's general condition slowly improved, and he was able to walk at the time of his discharge from the hospital in August.

In July, 1963, he was readmitted for elongation of the left Achilles tendon. A biopsy of the calcified area on the medial aspect of the left knee showed lamellar bone partly covered by fibrocartilage, with membranous ossification in progress at the boundary between the two tissues; no muscle was seen. On his last visit, he was walking well and the radiological evidence was unchanged.

Case 2. On July 1, 1962, a 32-year-old woman was admitted because of multiple injuries sustained when she fell from a moving car. She was fully conscious and had no signs of neurological injury. She required a tracheostomy and intermittent positive pressure ventilation because of a flail chest. She made good progress until July 8, when cardiac arrest occurred. The heart was readily restarted, but she remained unconscious for 3 months and had evidence of residual brain stem damage.

In October, she regained consciousness, but her limbs remained spastic; the legs were extended. X-ray films showed marked calcification around both elbows and knees (Fig. 2). The patient made no further improvement and died in the hospital 18 months later. The calcification remained unchanged.

Case 3. A 7-year-old girl was admitted to the hospital on May 27, 1966, after being hit by a car. She was unconscious at the time of admission and remained so for the next 14 weeks. The tone and reflexes were increased in all four limbs and she showed bilateral Babinski responses. Temperature elevations to 103°F were controlled by chlorpromazine and surface cooling.

In July, she was found to have hard bony swellings in relation to the shaft of the left humerus and both femora. X-ray films confirmed the presence of soft tissue calcification at these sites as well as a small amount of calcification around the hips. The serum calcium and phosphorus were normal, but the alkaline phosphatase was 24 King Armstrong units.
Barnett and Nordin\textsuperscript{2} have set forth a "femoral score" in such situations, which they define as "the sum of the thicknesses of the 'medial' and lateral cortices of the femoral shaft at the thickest point of cortex, divided by the total diameter of the shaft at that level. This fraction is multiplied by 100." Femoral scores on this patient were as follows: July 1966, left 38.5, right 37.5; April 1967, left 28, right 26.

By September, the patient was improving rapidly; she was then able to talk, although with difficulty. In May, 1967, she entered a school for handicapped children because of spasticity of the left arm and both legs. X-ray films taken in April, 1967, showed almost complete disappearance of the heterotopic calcification, which remained only as a spur on the shaft of the right femur (Fig. 3) and at the left elbow.

**Discussion**

The biopsy taken in the first patient showed bone but no muscle. The relationship between dystrophic muscle calcification and the ossification (myositis ossificans) that occurs in neurological disorders is very close. The essential factor in the pathogenesis of both is the mineralization of the connective tissue. Wolff\textsuperscript{16} has shown that the initial calcification occurs in relation to the binding of labelled protein, and it is possible that this corresponds to the initial phase of Urist's triphasic mechanism.\textsuperscript{13,14}

*Two Types of Muscle Calcification.* It is likely that there are two distinct groups of dystrophic muscle calcification. The first group due to localized trauma shows localized calcification around a joint which is involved in a muscle contracture. In a second group involving widespread calcification, a severe metabolic disturbance is of prime importance.

The formation of bone is usually ascribed to metaplasia of fibroblastic connective tissue elements, or to damage of periosteum with the liberation of osteoblasts. The anatomical distribution of the lesions is not...