MYOSITIS OSSIFICANS PROGRESSIVA

K. S. Grewal and N. Das, Amritsar, India

From the V. J. Hospital, Amritsar

Mather (1931) and Ryan (1945) affirm that this disease occurs exclusively in the white race. Our case, being in an Indian native child, is therefore of interest. So far only 161 cases of this rare disease have been reported in the world literature.

![Figure 1](image1.png)  ![Figure 2](image2.png)
Figure 1 - The child trying to abduct his shoulders and to open his mouth: the jaws cannot be separated. Both elbows ankylosed. Figure 2 - Generalised kyphosis, and a bony nodule on the back.

![Figure 3](image3.png)  ![Figure 4](image4.png)
Figure 3 - Microdactyly of great toes and hallux valgus of both feet. Figure 4 - The proximal phalanx of the hallux is dislocated laterally and lacks a basal epiphysis. This may perhaps be represented by the anomalous centre at the head of the first metatarsal bone.

CASE REPORT

A seven-year-old Indian complained of inability to open his mouth, and of progressive stiffness of the back and limbs since infancy. He was healthy till he was six months old,
when firm nodules appeared on the back and gradually increased in size. After another six months a fresh crop of nodules appeared about the shoulders. During the next year, all these nodules started to become smaller, but bony hard. In the last three and a half years the joints have become stiff; he cannot open his mouth, so that he is restricted to a semi-solid and liquid diet, and he has to be fed because his hands do not reach his mouth. Family history—A careful history failed to reveal any affection of relatives. Of two elder brothers, one has spinal caries.

Clinical examination—He was apparently a well built and well nourished child. The mouth could not be opened. The neck was short, stiff and immobile (Fig. 1). Respiration was diaphragmatic. Only the left side of the abdomen moved. There was a generalised kyphosis (Fig. 2). A continuous bony ridge was felt along the antero-lateral aspect of the whole of the right half of chest and abdomen, extending down to the right iliac crest. Upper Limbs—Gleno-humeral movements were absent, but there was slight mobility of each scapula on the chest. The right elbow was ankylosed at 120 degrees, and the left at 130 degrees. The wrists and hands were normal in shape, size and movements. Lower limbs—The right hip was fixed in flexion at 90 degrees. Left hip movements were normal. The knees were flexed, but movement was possible through 5 to 10 degrees. There was microdactyia of the great toes (Fig. 3) with halluc valgus. Ankle and foot movements were normal. Urine—Normal.

The patient was able to sit on a stool, but was unable to walk even with the help of crutches because his arm muscles were too weak.

Radiographic examination—Figure 5 shows bone formation in the muscles of mastication. Figure 6 shows the continuous bony ridge on the front of the right chest and abdomen. Figure 7 shows bone in the sternomastoids. Figure 4 shows shortening of each great toe; its proximal phalanx is dislocated laterally and is without evident basal epiphysial centre, but this may be represented by an anomalous centre of ossification adjacent to or fused by cartilage to the first metacarpal bone.
Pathological examinations. Blood—Total white cells 16,000 per cubic millimetre; polymorphs 42 per cent; lymphocytes 34 per cent; mononuclears 2 per cent; eosinophils 22 per cent. Serum calcium 10.6 milligrams per cent; inorganic phosphates 4.37 milligrams per cent. Alkaline phosphatase 3.23 units per cent. V.D.R.L. and Meniki negative. Stools—Repeated examination for parasitic infections was negative.

Operation—A piece was removed from the ridge of bone on the right abdominal wall. Bone formation recurred with added zeal.

COMMENT

The disease is characterised by generalised ossification of the skeletal muscles. Really it is fibrositis ossificans progressiva (Greig 1931) for muscle fibres are not primarily ossified. The cause is unknown. Biochemical investigations have shown no appreciable change either in the blood chemistry or in the internal secretion of ductless glands. Münchmeyer (1869) described the histology, which is well summarised by Frejka (1929).

Frejkat distinguished four types. 1) The usual type, slow and gradual in onset, occurs insidiously in early childhood: our case falls into this category. 2) A type that begins with inflammation of some sort: his own case and some others belong to this category. 3) A type beginning with trauma: new bone forms with each injury. 4) Bone formation after transverse lesion of the spinal cord (myositis ossificans neurotica)—a distinct condition.

Except in a singular case of Rosenstirn, in which ossification extended into the subcutaneous tissues, ossification remains confined to voluntary skeletal muscles, tendons and ligaments. The disease is progressive, though liable to some waxing and waning. The hands and feet escape and the rest of the body becomes bony stiff. Death occurs from intercurrent disease or from chronic starvation due to ossification of the muscles of mastication. Removal of the bony masses results in reappearance of new bone, often in increased amounts.

REFERENCES


