MYOSITIS OSSIFICANS PROGRESSIVA*
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This is a rare progressive disease of unknown etiology in which ossification occurs in tendons, ligaments and fascia in addition to the interstitial tissues of the muscle.

Gay Patin in 1692 described the first case of a woman "who turned into wood". Freke in 1740, and Copping in 1741 reported similar cases. With the advent of radiography, the diagnosis was more easily made. In 1868, Von Dusch gave the disease its name. Lutwak in 1964 reviewed 260 cases from the world literature and added 6 new cases of his own. To our knowledge only 6 cases have been reported in the Indian literature so far (Dikshit and Tandon 1967, Tuli and Sharma 1970). This prompted us to report this case.

Report of a Case

The patient was a 4-year-old Muslim boy, the fifth child of healthy parents. He was the product of a full term normal pregnancy. At birth it was noted that he had a deformity of the great toes (hallux valgus). When the baby was only 15 days old, the parents noticed a small swelling in the occipital region, which turned into a firm mass, by the time he was one year old. His early growth and development were normal and he had started walking by the age of 1½ years. Since then he had developed several firm swellings in the region of the posterior part of the neck, para-vertebral muscles, chest wall, muscles of the shoulder girdle and abdominal wall. The swellings were firm and tender at the onset, but later became hard nodules with a bony consistency. Within a year his neck became immobile. Similar swellings had appeared in the axillary folds and arms during the previous year, limiting the movements of shoulder and elbow joints, so that he had difficulty in feeding himself.

He had 4 normal siblings. There was no history of consanguinity or similar complaints in the family.

On examination, the child was a well nourished boy of average intelligence. The weight was 13.5 kg, the height 93 cm. He had a characteristic 'poker man' attitude with a stiff neck. There were hard bony swellings along the ligamentum nuchae, para-spinal muscles, serratus anterior, trapezius, lattisimus dorsi and abdominal wall muscles. The spine was rigid throughout. There was a restriction of movements at the shoulder and elbow joints. The forearm, hand and lower limbs were comparatively free. The movements at the wrist, hip, knee and ankle joints were normal. Hallux valgus deformity was present on both sides. There was difficulty in opposing the left thumb. The other toes and fingers were normal. The rest of the physical examination was normal.

The routine examination of the urine and blood was essentially normal.

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Serum calcium and phosphorus levels were within normal limits. X-rays of the skeletal system showed huge sheets of calcification on both sides of the vertebral column from the occiput to the pelvis, calcification of the ligamentum nuchae and muscles of the shoulder girdle.

A biopsy was taken from the lattisimus dorsi muscle. Histopathological examination showed sections of the muscle with areas of cartilage formation and ossification suggestive of myositis ossificans.

Discussion

The disease involves the connective tissue, but the exact mechanism is not known. The disease is familial. Cases have been reported to occur in 3 consecutive generations and in homozygous twins. 75% of the affected persons have other congenital abnormalities, like microdactyly or adactyly of the thumb or great toe, hallux valgus, polydactyly, deformed ears, abnormality of the teeth and spina bifida. Involvement of the great toe is the commonest. Digical abnormalities are always symmetrical. Less frequently maldevelopment of the gonads, cardiovascular and nervous systems have been described.

The age at which the initial symptoms appear varies considerably. The disease usually manifests itself during the first few years of life and almost always before the age of twenty years. Typical lesions have been found even at birth.

Without any apparent cause or after trivial trauma or infection, swellings appear in muscles, associated with heat and pain. The swelling slowly disappears leaving a firm mass in the region of the muscle, which later calcifies and eventually becomes bone. The sites frequently involved are the paravertebral and cervical regions, followed by muscles of the shoulder girdle, proximal musculature of the arm, the pelvic girdle, and muscles of the jaw and head, as in this case. The bony lesions are small at first and confined to the muscle. Later they coalesce and get attached to the bones of the skeleton and cause limitation of movements of the involved area. The neck and scapulae are first immobilised. Ultimately huge sheets of bone are formed over the trunk and neck. The hands, forearm, hips and legs are not usually involved. The heart, diaphragm, sphincters and laryngeal muscles are spared. Death is usually a result of respiratory embarrassment due to involvement of muscles of the thorax or due to inanition following involvement of the masseter and temporalis muscles.

No biochemical abnormalities have been reported. Serum levels of calcium, phosphorus and alkaline phosphatase are within normal limits. Urinary excretion of calcium and phosphorus is also reported to be normal. High values of alkaline phosphatase have been reported at biopsy.

Radiologically, the ossified areas appear at bony ridges within the tendon and muscle area.

Histopathologically the early lesions show swelling and proliferation of the connective tissue. The involved muscle fibrils show scanty nuclei and hyaline degeneration of the cytoplasm. In the region of calcification, the tendon and muscle is replaced by a collagen like material and deposits of calcium. When extensive ossification has occurred, the