FIBRODYSPLASIA OSSIFICANS PROGRESSIVA (A Case Report)

Lt Col SS ANAND*, Maj P JOSHI+

MJAFI 1995; 51 : 213-214

KEY WORDS : Fibrodysplasia ossificans progressiva.

Introduction

Fibrodysplasia ossificans progressiva (synonyms : Myositis ossificans progressiva and Munchmeyer's disease) is a rare inherited disorder with a prevalence of less than 0.1 per million population. It is characterised by extensive ossification of the connective tissue associated with typical skeletal abnormalities. The skeletal abnormalities are usually present since birth and may alert the radiologist to the subsequent development of fibrodysplasia ossificans progressiva.

CASE REPORT

A 5-year-old female child was brought to hospital with complaints of deformities of the feet since birth. The parents gave history of gradual onset of multiple bony swellings over the scapula, spine and humerus. They also gave history of the child's inability to move her neck and bend her spine. The child was an offspring of nonconsanguineous marriage. There was no significant antenatal history. The parents and two other siblings were normal. The child was of normal stature. There was no relevant finding on systemic examination. Local examination revealed hard swellings of various sizes over the neck, back and arms. Bilateral hallux valgus deformities were also present. Radiological examination showed evidence of soft tissue ossification over the posterior aspect of the neck extending from the occipital region to the lower cervical region (Fig 1). Extensive soft tissue ossification was seen posteriorly and laterally on a radiogram of the chest (Fig 2). Ossification in the paraspinal region was also seen in the chest and abdomen. The left elbow showed a fixed flexion deformity of 140 degrees. Pseudoexostosis formation was seen over the left humeral shaft. Both feet showed evidence of hallux valgus with absent distal phalanges of great fourth toes (Fig 3). A radiogram of the pelvis showed bilateral femoral neck widening.

Discussion

Fibrodysplasia ossificans progressiva has been recognised since the seventh century, although it has been labelled by a variety of names including myositis ossificans

Fig. 1 : Radiograph of the cervical spine (lateral view) showing large posterior arches with ossification in the soft tissues of the neck.

Fig. 2 : Chest radiograph showing ossification in the soft tissues of the chest wall bilaterally.

*Classified Specialist (Radiology and Nuclear Medicine) Military Hospital Nankum, Ranchi; +Radiologist, On study leave, Ruby Hall Clinic, Pune 411 001.

Fig. 3 : Radiograph of the feet showing bilateral valgus deformities and malformations of the phalanges of the great toe and the fourth toe.

progressiva. Fairbanks [1] suggested the name fibrodysplasia ossificans progressiva as the ossific deposits are laid down in the connective tissue between muscle fibres and not in the muscles. The onset of the disease is usually before the age of five years with a male preponderance [2]. In our patient the symptoms started at the age of two years and had been progressing gradually ever since. The deformities of the feet were present since birth. Deafness has been seen in almost a quarter of all patients [3] but was absent in our case.

The characteristic radiographic features consist of soft tissue ossification which are seen as thick irregular bands or sheets in the neck, trunk and proximal limbs. Ossification of ligamentum nuchae is often seen and acts as a strut between the occiput and neck [4] as was also seen in our patient.

Hip joints reveal acetabular dysplasia with apparently enlarged femoral heads and necks. Dysplasia of the mandibular condyles with ankylosis of the temporomandibular joints has also been reported [5]. Widening of the femoral necks was seen in our patient too.

The typical feature in the hallux necks was seen in our patient too. The typical feature in

the hallux is an abnormality of the first metatarsal either in the form of pseudoepiphysis or distal medial protuberance, with either a segmental abnormality or synostosis [4] as was seen in our case.

Bone scanning and computed tomography (CT) have been used to monitor the progress of disease [6]. Bone scan is more sensitive than plain radiography in monitoring the full extent of involvement while CT can demonstrate small foci of calcification within the connective tissues and also flattened sheets of ossification around the muscles.

It is now well established that trauma, including surgery, may aggravate the condition and induce massive ossification [7]. Radiologists thus play an important role by suggesting the correct diagnosis and restricting biopsy and other surgical procedures. The late sequelae are respiratory complications because of the rigidity of the chest wall and inanition due to temporomandibular joint ankylosis.

REFERENCES

1. Fairbanks HAT. Myositis ossificans progressiva. *J Bone Joint Surg* 1950; 32B : 108-11.
2. Cramer SF, Ruehl A, Mandel MA. Fibrodysplasia ossificans progressiva : A distinctive bone forming lesion of soft tissues. *Cancer* 1981; 48 : 1016-8.
3. Connor JM, Evans DAP. Fibrodysplasia ossificans progressiva : the clinical features and natural history of 34 patients. *J Bone Joint Surg* 1982; 64B : 76-83.
4. Thickman D, Bondadapour A, Clancy M, Steel H. Fibrodysplasia ossificans progressiva. *Am J Radiol* 1982; 139 : 935-41.
5. Renton P, Rarkins SF, Stamp TCB. Abnormal temporomandibular joints in fibrodysplasia progressiva. *Br J Oral Maxillofac Surg* 1981; 19 : 318-25.
6. Carter SR, Davies AM, Evans N, Grimer RJ. Value of bone scanning and computed tomography in fibrodysplasia ossificans progressiva. *Br J Radiology* 1989; 62 : 269-72.
7. Rogers JG, Geha WB. Fibrodysplasia ossificans progressiva : A survey of 42 cases. *J Bone Joint Surg* 1979; 61A : 909-14.