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MYOSITIS OSSIFICANS PROGRESSIVA IN A NIGERIAN INFANT

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SUMMARY

Myositis ossificans progressiva, a progressively destructive musculoskeletal system disease is a well-described entity amongst Caucasians.

A case report, documenting its early manifestation and associated features in a Nigerian infant is presented here.

Key Words: Myositis Ossificans Progressiva, Infant Nigeria.

INTRODUCTION:

Myositis ossificans progressiva, otherwise called Munchemeyer's Disease, is rare, though well documented.[1]. It is a variant of a group of diseases in which there is metastatic calcification in man.[2] This group of diseases has two varieties viz: the first is present when there is a scattered amorphous deposit of calcification (e.g hypervitaminosis D) and the second occurs when there is true osseous tissue[2]. In the latter group are myositis ossificans traumatica and circumscripta (following trauma or infections e.g. poliomyelitis) that tend to remain localized, and myositis progressiva, a generalized, and presumably, a systemic disorder which is the subject of this paper.

It is characterised by the formation of areas of calcification in the interstitial connective tissue of muscles, tendons, ligaments, fascia and aponeuroses,[3] with eventual replacement of these structures with columns and plates of bone[1]. Though its first sign of occurrence is often heat, oedema and pain in a muscle, often starting with the sternocleidomastoid and then spreading caudally from the shoulder girdle to the pelvic girdle,[1] the ultimate effect however is an insidious loss of body motion[3].

This disease is thought to be a disease that manifests congenitally or early in life and in which there are associated congenital anomalies of digits, often expressed as microdactyly or adactyly of the thumbs and the great toes.[2]

A report of its occurrence in a Nigerian child is presented here.

CASE REPORT:

C.U., a 7 month old male presented to us on referral with a history of soft tissue swellings on the upper back region and neck that were on and off (with or without treatment) from the 8th day of life. Other problems included transient depressions on the dependent side of the head on waking up that was noticed from 3 months of life, at 4 months, the swellings on the sides of the chest persisted and became hardened with a resultant positioning of the upper limbs in a hanging posture. At 5 months of age, both sides of the neck became swollen and painful when touched but this resolved after 2 weeks but had a residual effect of restricting the child's head movement to the left. There were however no feeding or respiratory difficulties. All other histories were not significant. Their family is a monogamous one and his siblings do not have bodily or limb deformities.

He was found on examination to be well nourished with the shoulder joint in semi extension and abduction in addition to the torticollis of the left sternocleidomastoid muscle.

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His sclera was bluish and there was pallor but no cyanosis or jaundice. He had features of craniotabes over the parietal bones. He was also edentulous. His weight was 9.4kg and his head circumference, 48cm.

Musculoskeletally he had microdactyly of the halluces of both feet (fig. 1) and he had hard fixed masses spanning from the latero-inferior border of the scapular to the lateral surfaces of the 5th, and 7th ribs on both sides. The blood count revealed a hematocrit of 27%, total white cell count of 6.4* 10^9/L with a differential count of 25% neutrophils, 67% lymphocytes, 5% eosinophils and 3% monocytes. The erythrocyte sedimentation rate was 11mm/1stHr (Westergreen). His blood chemistry showed; calcium, 2.6mmol/L (normal range, 2.2-2.8mmol/L); inorganic phosphate, 1.6mmol/L (normal range 1.1 - 1.6mmol/L); alkaline phosphatase 148iu/l (normal range 25 - 210iu/l for children).

Roentgenogram of the skeletal system done was as follows;

Fig II Chest Exuberant soft tissue calcifications both axillae, the lateral thoracic areas and along the humeri.

Fig III Both hands. Shortened 1st metacarpals both hands, clinodactyly left little finger.

Fig IV Both feet Shortened 1st metatarsals bilaterally. Bilateral halux valgus deformity.

He was started on prednisolone 10mg 12 hourly for 3 weeks but was lost to follow-up.

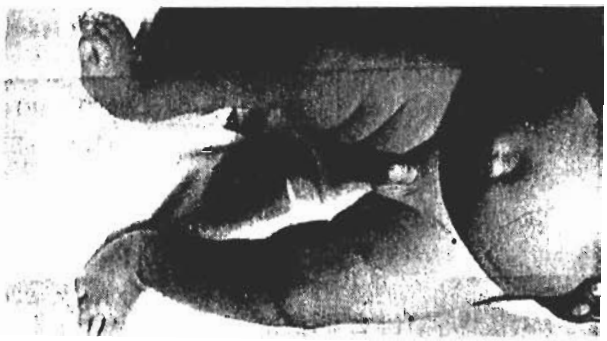

Fig. 1 - Microdactyly of Halluces of both Feet

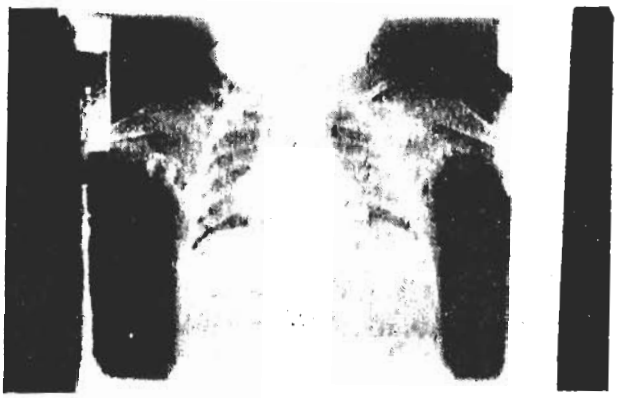

Fig. 2. Exuberant Soft tissue calcifications Both Axillae, The lateral Thoracic areas and along the Humeri.

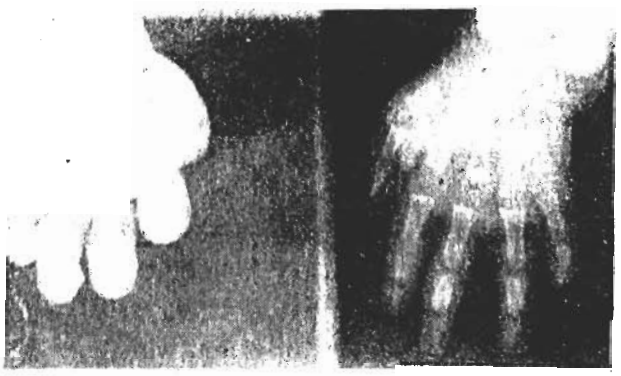

Fig. 3 - Shortened 1st Metacarpals both hands; Clinodactyly left little finger

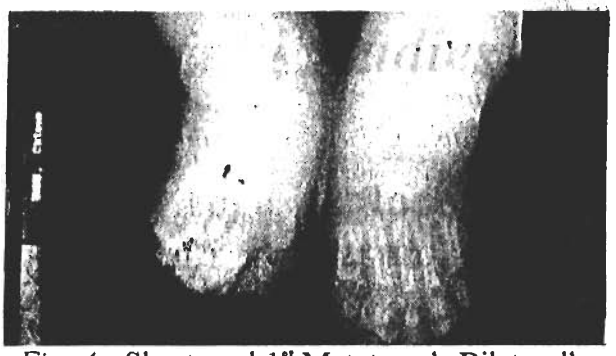

Fig. 4 - Shortened 1st Metatarsals Bilaterally; Bilateral Hallux valgus deformity

DISCUSSION:

Most of the currently available literature on this disease is from developed countries, afflicting Caucasians mainly. This case report involving a Nigerian infant has tried to document some early features of the disease as seen in our patient. Though the biopsy of the mass was not done, the features on this child at presentation were as found in the disease. As documented by other authors,[12] the swellings on this patient tended to relapse and resolve over time, involving different sites and affected mainly the upper part of the back, the sides of the chest and the neck. These swellings however became permanent and more hardened from 4 months onwards.

The other features on this patient included the bluish sclera and the craniotabes that were noticed at 3 months of life.

We could not follow up the patient to determine the impact of the steroids we gave to him.

This case is being reported to highlight the occurrence of this disease in a Nigerian and the fact that if patients are well assessed, it could be picked early.

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