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Research Article

LOCALIZED GIGANTISM: SERIES OF 15 CASES

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ABSTRACT

The Study Of 15 cases of Localized form of gigantism in different congenital conditions was carried out in Mmcri, Mysore, Kims Hubli, and Esic Kalaburagi during The Last 30 years. Here patients werre mainly seeking medical advice for cosmetic reasons & for disability certificates. We took clinical photographs, plain radiographs of affected limb, USG (Gray scale & Doppler-showed soft tissue thickening & no Increased blood flow).& biopsy of tissue. Some patients refused for the same. In all the above cases there was abundance of fibrofatty tissue along with dense fibrous tissue, in few cases especially upper limb, nerves were enlarged &thickened. It was confined to
MEGAFOOT- 4 Cases Since Childhood, Stretched Shiny Scaly Dry Skin, Double The Girth Of Opposite Limb Syndactyly (2 & 4TH) TOE, Rudimentary(1ST& 5TH TOE Variation)
Unilateral giant lower limb -2cases
Giant upper limb- 3 CASES
Radial Mega Hand
Mega Thumb-3 cases
Macrodactaly of Index Finger etc……
It Was Compatible With Normal Life.
We Concluded that it is a rare type of disease. No genetic involvement, any theories cannot be formred or dimissed. Few were associated with Neurofibromatosis.
HPE**:** Excessive Proliferation & Accumulation of Fat is the basic lesion. Surgical correction is the treatment of choice.

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INTRODUCTION

Localized Gigantism is described as a rare, non-hereditary, congenital condition presenting with localised macrodactyly and a proliferation of mesenchymal elements.[1]There is in particular a marked increase in fibroadipose tissue.[1]It usually involves areas of distribution of the plantar and median nerves[2]. The localized gigantism is almost invariably recognized at birth, but starts to cause problems as the child grows. There may be difficulty in walking and in performing routine activities, but cosmetics are the main issue concerning the patient requiring consultation with surgeons. This disease is usually unilateral. The growth velocity may differ from digit to digit. The lower limb is more frequently involved than the upper limb. The abnormal area is usually along a specific sclerotome. The second and third digit of the hand and feet are most frequently involved, corresponding to the median nerve and medial plantar nerve supply in the upper and lower limb[3,11.]

MATERIAL AND METHOD

We had an exceptional opportunity to study 15 cases of localizedgigantismin the department of orthopaedics MMCRI MYSORE, KIMS HUBLI, And ESIC KALABURAGI, during The Last 30 Years. Here patients were mainly seeking medical advice for cosmetic reasons & for disability certificates. We took clinical photographs, plain radiographs of affected limb, USG (gray scale &doppler)-showed soft tissue thickening (fig:1) & no Increased blood flow.& biopsy of tissue. Some patients refused for the same. Out of these Four were of mega foot and two cases giant lower-limb involvement, three were giant upperlimb cases, three were macrodactyly of the thumb and 1 case of radial megahand, over a period of 30 years. Although parents noted the abnormalities shortly after birth, none of the patients presented before five years of age. Increasing difficulty in wearing shoes and cosmetic worries were the main cause for seeking medical advice. The presenting age ranged from 8 to 45 years. Nine of these were male while six were female (3:2). A detailed history revealed that the enlargement began shortly after birth in all the cases and the affected limb or digit grew at faster rate.

Clinical examination of the affected limb or finger or toe revealed thickened pale, glossy and non-tender skin. Consistency was firm in some areas and soft in others. Dorsal and lateral curvature of the affected digit was seen in two cases,

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while plantar flexion was seen in one case. One of the patients had associated neurofibromatosis. None had any other associated congenital anomalies. No area of pigmentation was noted anywhere. Chromosomal studies performed demonstrated no abnormalities in any patient. None gave a family history of any such deformity. Clinical photographs and radiographs were taken in every case. In five cases de-fatting surgery was initially undertaken, but due to recurrence of the deformity, amputation of varying degree was required in all the cases. Inability to remove all the fibro-fatty tissues combined with subsequent re-growth was the main reason of failure. All the excised specimens were examined histologically. (fig:4) Table I shows the relevant details of all cases. Ultrasound (Gray scale and Doppler) (fig:1) examination of the digits revealed diffuse soft tissue thickening. No evidence of increased blood flow was found in the affected region in any of the cases.

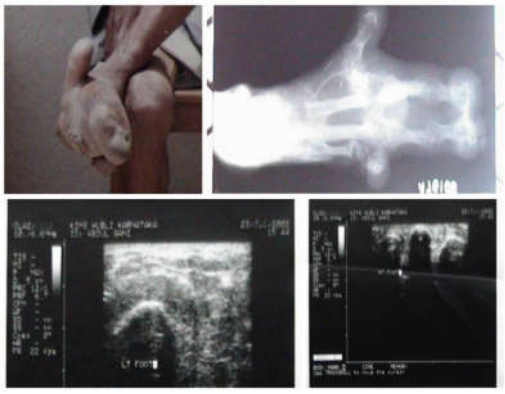

Fig 1 showing mega foot of male patient Clinical photograph Xray sonography

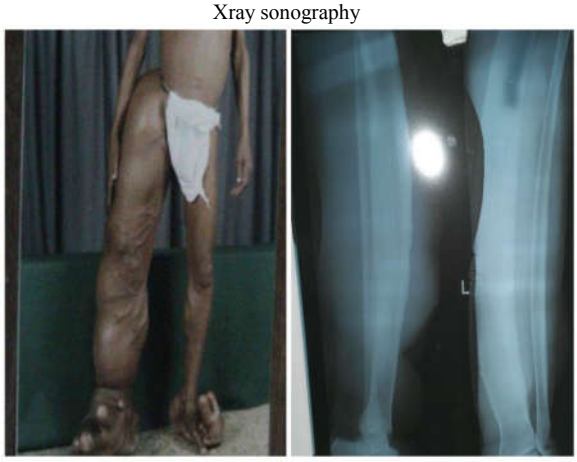

Fig 2 showing Male Patient with Giant Lowerlimb Clinical Photograph Xray

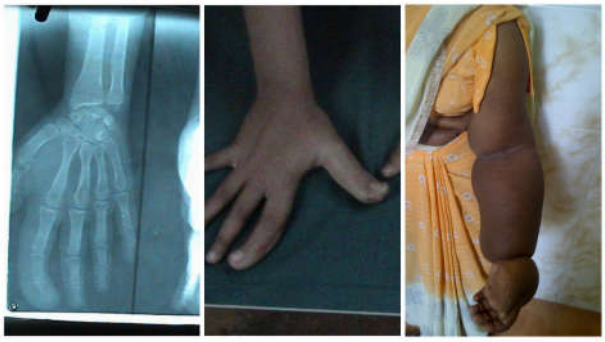

Fig 3 Showing Xray of mega thumb and Index Finger With Clinical Photograph Giant Upperlimb of Female Patient

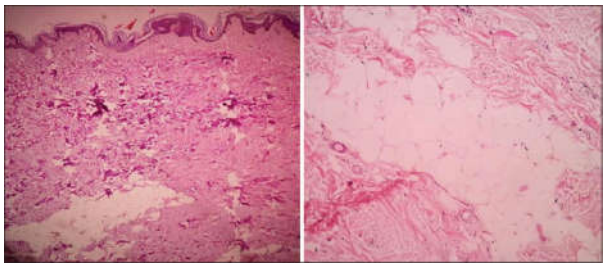

Fig 4 Histopathological Examination Abundant overgrowth of fibro-fatty Tissue The Hypertrophied Adipose Tissue With Very Large Lobules of Fat Pervaded All Surrounding Tissues

RESULTS

Table 1

Case no	Age(years)/sex	Location of anomaly	Digit involved	Syndactyly[10]	Nerve involvement	Associated conditions
1.	45/male	Foot	All five toes	*Present*	No	None
2.	8/male	Hand	Second and thumb	Absent	No	None
3.	26/male	Lowerlimb	Whole limb	Absent	No	*Neurofibromatosis*
4.	16/male	Foot	All five	Absent	No	None
5.	8/female	Hand	Thumb	Absent	No	None
6.	40/female	Upperlimb	All five	Absent	Yes	None
7.	25/male	Lowerlimb	All five	Absent	No	None
8.	21/male	Upperlimb	All five	Absent	Yes	None
9.	12/female	Foot	All five	Absent	No	None
10.	28/male	Foot	All five	Present	No	None
11.	11/female	Hand	Thumb	Absent	No	None
12.	29/male	Hand	Second	Absent	No	None
13.	23/male	Hand	Thumb	Absent	No	None
14.	16/female	Hand	Radial side	Absent	No	None
15.	44/female	Upper limb	All five	Absent	No	None

Differential diagnosis

Table 2

Increase in all mesenchymal elements	True Macrodactyly (MDL)
Overgrowth of a single element	Haemangioma Lymphangioma Fibrolipomatosis Congenital neurofibromatosis Proteus syndrome Klippeltrenauney- weber syndrome Idiopathic localised gigantism

DISCUSSION

In this study, we found that localizedgigantism occurs primarily before the age of eight. Problems start to surface with the growth of the child. Toddlers are reported to have difficulty in wearing shoes and to sustain repeated injuries, which may affect their daily activities, especially their learning development, social interaction, and self-confidence .[4]

The literature indicates a male preponderance .[5,6,7] The male to female ratio in this series was 3:2. None of our patients had any family history of similar deformities. This is consistent with the previous literature, which states that heredity does not play a role .[5,7,8]

The etiology of the localized gigantism remains unclear. As the child grows there may be degenerative changes of small joints and compression of neurovascular structure. Histopathologically, cut sections are rich in adipose tissue sprinkled in a fine lattice like fibrous tissue as in our case[9].Plain radiographs demonstrate abnormalities in both the soft tissues and bony elements. The affected long bones ,phalanges and metatarsal bones show an increase in width and length, and are often splayed at their distal ends, giving a mushroom like appearance. The articular surface may slant and in late childhood, severe secondary degenerative changes may affect the joints[3]. Ultrasound can assess the soft tissue and changes in the nerve. These are better visualized on MRI. A CT is better to evaluate the changes in bone. The differential

diagnosis of localized gigantism includes Macrodystrophialipomatosa(MDL), Lymphangiomas, Hemangioma, Klippel-Trenaunay-Weber Syndrome, Neurofibromatosis, Proteus Syndrome, and Fibrolipomatosis[2] .Surgical correction is the treatment of choice. The main surgical principle in treating this condition is to improve the cosmetic appearance, and to preserve the neurological function as far as possible[6] .Surgery is usually carried out after puberty when growth ceases. Cautious and planned use of multiple Debulking procedures, Epiphysiodesis, and various Osteotomies are recommended to achieve the best results. However, complications associated with overzealous debulking procedure lead to nerve injury with an incidence reported as high as 30-50%[14]. A localized recurrence rate of 33-60% makes the management of localized gigantism more demanding.

CONCLUSION

Numerous aetiologies of local gigantism exist. While clinically the distinction may be difficult, radiological investigation is very useful in confining the differential diagnosis.

A proper clinical examination, plain radiography, and ultrasound can diagnose localized gigantism confidently, thus obviating the need for MRI, especially in a poor socio-economic set up. Amputation with prosthesis can be performed with satisfactory results if the gigantism is very huge and the patient is not ready for multiple debulking surgeries.

Considering the rarity of disease and in absence of any genetic involvement, no theory can be easily formed or dismissed. Few Were Associated With Congenital Neurofibromatosis. Our histopathologic findings suggest that excessive proliferation and accumulation of fat is the basic lesion, whereas in macrodactyly of the hand involvement of nerves might be the fundamental lesion[13]. In addition to our study, following careful review of the literature, we noted that the selectivity of the disease is such that only a single digit is affected, or when more than one digit is affected, usually these digits are adjacent. Furthermore, we observed overgrowth of the surrounding tissues even after ablative surgeries.

***Diferential Diagnosis*: Localised Gigantism**

- Fibrolipomatosis
- Congenital neurofibromatosis
- Proteus syndrome
- Macrodystrophic lipomatosa
- Klippel trenauney - weber syndrome
- Idiopathic localised gigantism

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