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# CONGENITAL ISOLATED GIGANTISM OF GREAT AND SECOND TOE- A CASE OF NON SYNDROMIC NEUROLIPOMATOSIS

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**ABSTRACT**
Local gigantism is a rare and curious medical anomaly. Most reported cases were due to lipomatosis and less frequently AV malformations. Neurolipomatosis as a cause of local gigantism is scarcely reported. To report and record 1st case of neurolipomatosis causing local gigantism. Two year old male presented with gigantic great toe and 2nd toes. They were larger, longer and thicker than the rest of the foot. MR evaluation confirmed hypertrophy, hyperplasia of soft tissue and hyperplasia of phalanges of 1st & 2nd toes. Surgical correction resulted in satisfactory foot making the child`s kinesiology functional.

## INTRODUCTION

Localized gigantism either of limb or a part of the body has been a point of curiosity for both common man and medical profession. Localized gigantism has been described affecting limbs most commonly due to vascular abnormalities AV fistulas, syndromic lipomatosis are other common factors [1,2,3]. Lipomatosis causing localized gigantism has been published elsewhere but predominant neural component in neurolipoma is scarcely published. Hence this attempt to report a case of congenital isolated localized gigantism

## AIMS AND OBJECTIVES

To place a record of congenital localized gigantism due to neurolipomatosis

## CASE DETAILS

2 year boy was brought with a giant great toe. He was a product of full term normal delivery. He was born with a big great toe that has grown 10 times in 2years of age. Child was able to walk with difficulty as great toe constituted 4 times in length and 5 times in breadth with rest of foot. Examination of proximal limb did not reveal AV fistula or café au lait spots or swellings elsewhere in the body. Gigantism primarily localized to the 3 phalanges of great toe and second toe. The phalanges were hypertrophied and hyperplastic and were 3 times of normal phalanges. There was soft tissue component more on the plantar aspect then the dorsal aspect MR evaluation revealed neural, fat components and gigantic phalanges of great and second toes

Child was taken up for a staged excision; in stage 1 gigantic great toe and second toe were excised. The foot was brought to the shape and size of opposite foot SANS great toe and second toe. This constituted removal of 95 % of gigantic neurolipomatic tissues; 6months later rest of the soft tissue was excised and advised custom made shoes. One year follow up child was running and walking well. Histopathology showed neurolipomatous tissue with significant neural tissue in center the gigantic bones that was otherwise unremarkable.

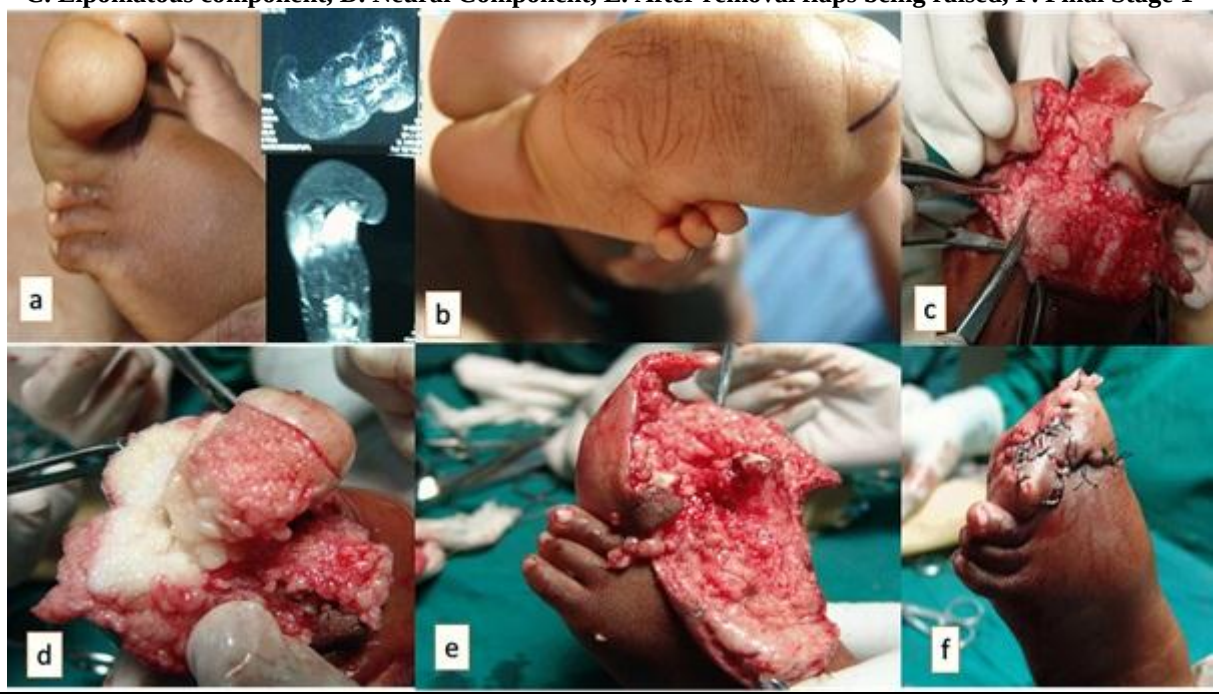
## DISCUSSION

Localized gigantism has been described anecdotally in literature most published reports [1-5] have

all been reported secondary to lipomatosis involving either entire or a part of the limb termed as macro dystrophia lipomatosa. In this index case the pathology was neurolipoma rather than a pure lipoma. The pathogenesis of gigantism and mechanism is ill understood. Local release of growth factors [6], increased blood supply in AV malformation are suggested pathomechanisms.

Difficulty in ambulation and walking, cosmetic disfigurement and progressive growth are primary indications of surgical intervention. Primary issue of surgical intervention is skin closure. Localized distal lesions were treated with amputation, staged excision, use of distant flaps for skin closure. Index case more than 95% of the neurolipomatous tissue was possible because lesion confined to great toe and second toes. The issue was creating a functional forefoot this was supplemented by use of custom made shoes.

**Figure 1. A. Clinical Picture showing Giant great toe, B. Plantar Gigantism more than dorsal component, C. Lipomatous component, D. Neural Component, E. After removal flaps being raised, F. Final Stage 1**




## CONCLUSION

Rare case of congenital isolated localized gigantism due to Neurolipomatosis successfully treated with surgery is being reported.

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