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E-ISSN : 2456-1045

- International Journal
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RESEARCH JOURNAL

VOLUME - 75 | ISSUE - 1

ADVANCE RESEARCH
JOURNAL OF
MULTIDISCIPLINARY DISCOVERIES

JULY
2022



INTERNATIONAL JOURNAL FOUNDATION

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Macroductyly- A case report

ORIGINAL RESEARCH ARTICLE

NAME OF THE AUTHOR(s)

ISSN : 2456-1045 (Online)
 ICV Impact Value: 74.80
 GIF- Impact Factor: 5.194
 IPI Impact Factor: 3.56
 Publishing Copyright @ International Journal Foundation
 Article Code: MDS-V75-I1-C2-JUL-2022
 Category : MEDICAL SCIENCE
 Volume : 75.0 (JULY-2022 EDITION)
 Issue: 1(One)
 Chapter : 2 (Two)
 Page : 05-07
 Journal URL: www.journalresearchijf.com
 Paper Received: 17th AUG 2022
 Paper Accepted: 24th AUG 2022
 Date of Publication: 10th SEPTEMBER 2022
 DOI: [10.5281/zenodo.7066422](https://doi.org/10.5281/zenodo.7066422)

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INTRODUCTION

Macroductyly is a rare non-hereditary congenital anomaly of finger and toes and it is very difficult to treat this, as it largely depends on exposure of surgeon to the disease process. The disease process may be static or progressive. It accounts for about 1% of upper extremity congenital anomalies. Usually involve fibrofatty tissue and sometimes bones. The first detail study was done by Barsky in 1967. It is linked with mutation in the PIK3CA gene which control cell proliferation and migration. Barsky defined it as an increase in size of all the elements or structures within a digit or digits¹. Kelikian coined the term "Nerve territory-oriented macroductyly" due to involvement of area along the digital nerve². This is an isolated case as no other deformity is present to group it under congenital deformity syndrome.

KEYWORDS: Macroductyly, congenital anomaly, fibrofatty tissue, congenital deformity syndrome.

CITATION OF THE ARTICLE



Gupta SK ; Singh S; Singh K ; Maurya V. (2022) Macroductyly- A case report"; *Advance Research Journal of Multidisciplinary Discoveries*; 75(2) pp.05- 07

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I. CASE REPORT

A 30-year-lady presented with congenital large finger of left-hand involving middle, ring, little finger and palm. The lesion is painless and gradually progressive. There is no significant history of any injury or surgery, but due to this condition her daily physical activity got progressively deteriorated. (Figure 1,2). Physical examination reveals an increase in the size of the fingers and palm. During complaint-directed history taking, the patient denied feeling pain but reported that the condition affected her daily activities and it doesn't look good. Frontal, oblique and lateral radiographs of the hand were requested, which revealed enlargement and deformity of the third, fourth and little fingers (figure 1,2). Diagnosis of macrodactyly was confirmed through clinical and radiological features.

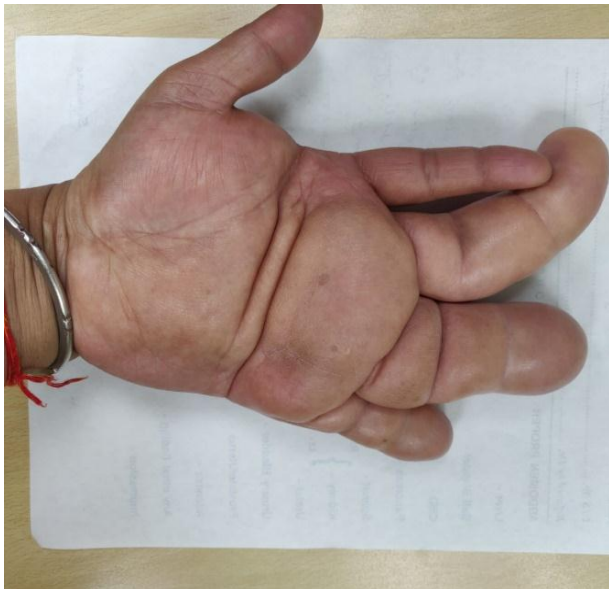


Fig-1 : Preoperative lesion



Fig-2: Preoperative lesion

II. INVESTIGATION

Xray findings- there is noted enlargement of the 3rd, 4th, 5th digits with associated enlargement of her phalanges. No soft tissue calcification or osseous mass seen. F/S/O Macrodystrophia lipomatosa Figure (3,4).



Fig-3 : X-ray hand AP view

Ultrasonographic findings- High-resolution ultrasound shows diffuse fatty proliferation in subcutaneous plane in middle 4th and little finger without any arteriovenous malformation.

Magnetic resonance imaging (MRI) scan- it shows soft tissue growth with prominent fibrous stand along phalanx with osseous hypertrophy of involved phalanx. These findings led to a strong suspicion of Macrodystrophia lipomatosa.

III. SURGERY

The main aim of surgery is debulking of fatty tissue with preservation of neurovascular bundle. Surgery is performed under TIVA. We chose to intervene in two stages, giving due consideration to the concern and choice of patient as during counseling

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patient got worried about loss of digit due to vascular compromise. Hence in the first stage, we plan to debulk the palm area leaving the fingers to be addressed during the second sitting. We give incision over palm in inverted "T" shaped and zig zag pattern to remove extra fatty tissue of palm with preserving important neurovascular bundle. And send specimen for histo-pathological examination (Fig. 3). Second stage of surgery is planned later on where we would remove extra fatty tissue from phalange's at that time we also plan for corrective osteotomy. During post op period given antibiotics and analgesic with great wound cared also advise flexion, extension exercise for interphalangeal joint.



Fig -4 intraoperative



Fig-5 post op after 1 week

IV. DISCUSSION

Macroductyly incidence in hand and feet is near about similar but it is seen that more commonly it affects along the median nerve³. Male preponderance is seen. Macroductyly surgery have not ensured complete response but helpful in respect of function and cosmetic purpose⁴. In between surgeon and patient should maintain good relationship and it should be fully explained what will be probable outcome of surgery. About amputation always think twice try to avoid debilitating surgery.

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