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Fibrolipomatous hamartoma emerging from the median nerve associated with macrodystrophia lipomatosa

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ABSTRACT

Fibrolipomatous hamartoma with macrodystrophia lipomatosa is a rare congenital condition characterised by a benign enlargement of the peripheral nerves and surrounding fibro-adipose tissues. A 7-year-old child was referred to our plastic surgery clinic with complaints of a massively enlarged right middle finger, a moderately enlarged index finger and a puffy hand. He had palpable, nontender enlargements of the fibro-adipose tissues (13 × 6 × 4 cm and 8 × 4 × 2 cm) extending to the palm and the wrist. They were slowly growing and had been noticed since birth. The diagnosis was based on magnetic resonance imaging. Functional and cosmetic considerations are important in such cases, and multiple debulking surgeries are often required.

Keywords: Macrodystrophia lipomatosa, Fibrolipomatous hamartoma, Median nerve, Enlarged finger, Exeresis.

1. INTRODUCTION

Neural fibrolipoma, also known as fibrolipomatous hamartoma, is an infrequentcongenital condition mostly seen in young patients. It is characterised by a benign augmentation of fibro-adipose tissue between nerve bundles, which results in overgrowth of the affected nerve (Chiang et al., 2010). The condition does not onlyaffect the branches of the median nerve but any peripheral nerve can be involved, and usually targeting the middle and index fingers. Macrodystrophia lipomatosa is a rare benign exaggeration of mesenchymal and fibro-adipose tissue that influences one or more digits on the distal upper or lower extremities, manifesting as a configuration of gigantism (Brodwater et al., 2000). These conditions co-exist in 40% of cases.

2. CASE PRESENTATION

A 7-year-old child with Down syndrome presented to our institution with a massively enlarged middle finger and a moderately enlarged index finger of the right hand, associated with a puffy hand and reduced mobility. There was no ecchymotic or petechial stain, skin defect or pitting oedema over the

affected fingers. The growing was steady, had been noticed since birth. It was associated with numbness and intermittent pain and had increased in severity over time. Magnetic resonance imaging (MRI) confirmed the diagnosis of macrodystrophialipomatosa. The decision was made to amputate for functional and cosmetic purposes but the parents refused permission to amputate both enlarged fingers at the same time. Therefore, the middle finger was scarcely amputated.

The patient presented at the age of 9 years old with massive extended enlargement of the right index finger. Upon examination, the patient was vitally stable. He had palpable, painless growing tissues of the right index finger extending to the distal phalangeal palm. There was no skin stain or defect and oedema over the enlargement. Movement was minimal and inadequate at the proximal and distal interphalangeal joints of the first finger but normal at the metacarpophalangeal joint. Scarring from the previous surgery was present. No sensory or motor deficit was noted (Fig. 1).

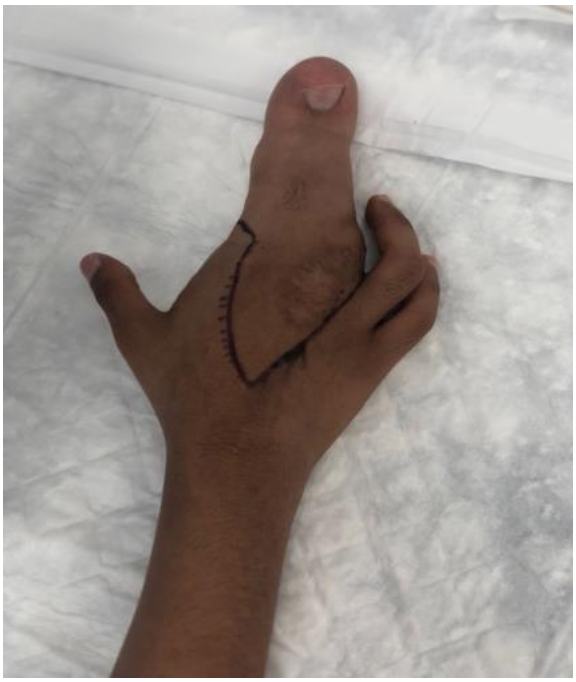

Figure 1 Macrodystrophia lipomatosa of the right index finger.

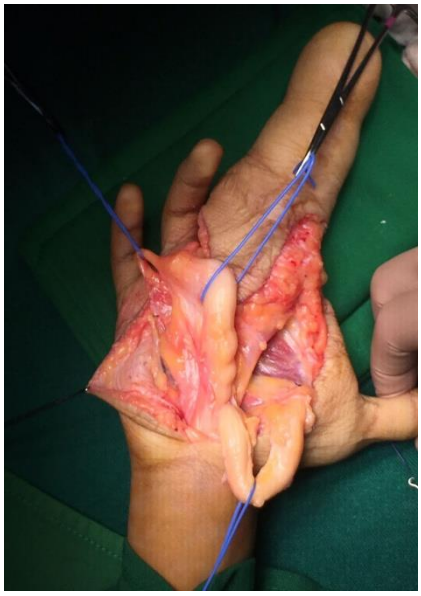

Figure 2 Fibrolipomatous hamartoma of the median nerve.

Under general anaesthesia, a ray incision was made around the right index finger. The median nerve branches were preserved to the thumb and ring finger via proper dissection and identification (Fig. 2). Upon incision of the flexor retinaculum, a copious

amount of adipose tissue was seen along the course of the median nerve. Amputation was performed by cutting off the index finger, and homeostasis was secured by tourniquet release to preserve perfusion. Skin closure was by Vicryl horizontal mattress suturing. Histopathological biopsy confirmed that the lesion was a neural fibrolipoma. The patient tolerated the procedure well and was seen multiple times after the operation (Figs. 3 and 4).

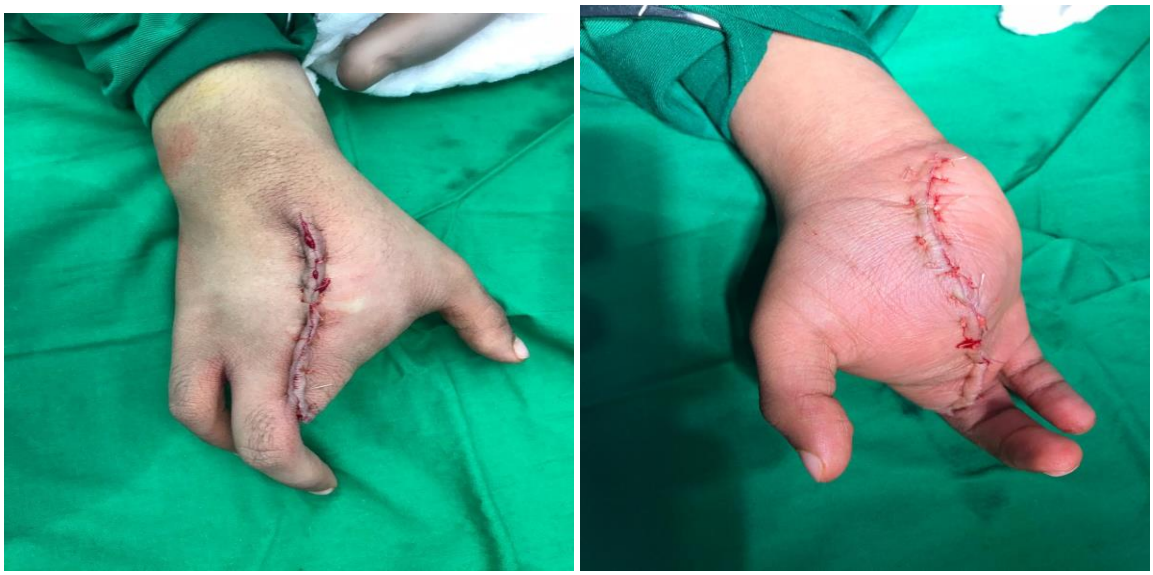

Figure 3 Patient's right hand after amputation of the enlarged index finger.

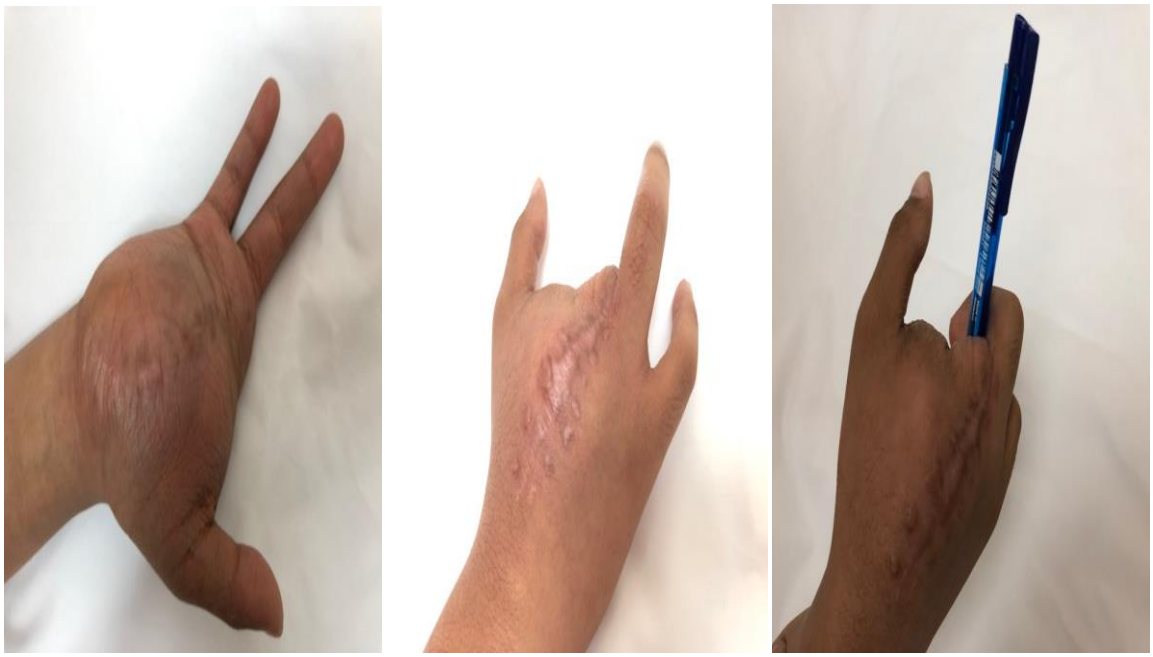

Figure 4 Patient's right hand after 3 months of amputation.

3. DISCUSSION

Macrodystrophia lipomatosa is a congenital non-hereditary macrodactyly distinguished by hyperplasia of fibro-adipose tissue and often coincide with periosteal and endosteal new bone genesis. Fibrolipomatous hamartoma can involve any peripheral nerve (Evans et al., 1997). Overall, it is mostly seen in the legs, with a preference for the plantar nerve. In the hands, it is frequently associated with the median nerve (Louaste et al., 2011). When such association occurs, it begins at the carpal tunnel and expands distally to the distal phalangeal palm. Nonetheless, participation of the proximal part of the median nerve above the wrist is unusual (Toms et al., 2006).

Sections revealed skin covered by a thick layer of keratin. The underlying dermis was fibrotic. The abundant well-delineated subcutaneous adipose tissue was surrounded by a thin fibrous network (Figs. 5 and 6). The bone marrow and skeletal muscles were

also infiltrated by adipocytes, and nerve hypertrophy was also seen. Fibrolipomatous hamartoma with macrodystrophia lipomatosa is uncommon. Radiographic and MRI hallmarks hold the promise of avoiding unnecessary biopsies and may lead to accurate diagnosis. In our patient, a plain radiograph showed bony hypertrophy resulting in a large right index finger or focal gigantism (Fig. 7). There was mild medial angulation of the second metacarpal digit over the proximal phalanx and absence of the third phalanx with abnormal soft tissue proliferation surrounding the second digit. The rest of the osseous structures appeared normal.

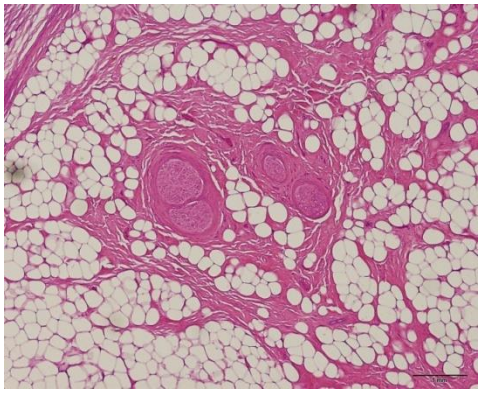

Figure 5 Nerve trunk hypertrophy

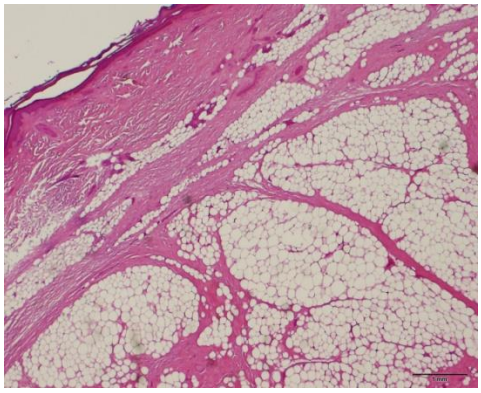

Figure 6 Increased subcutaneous adipose tissue.

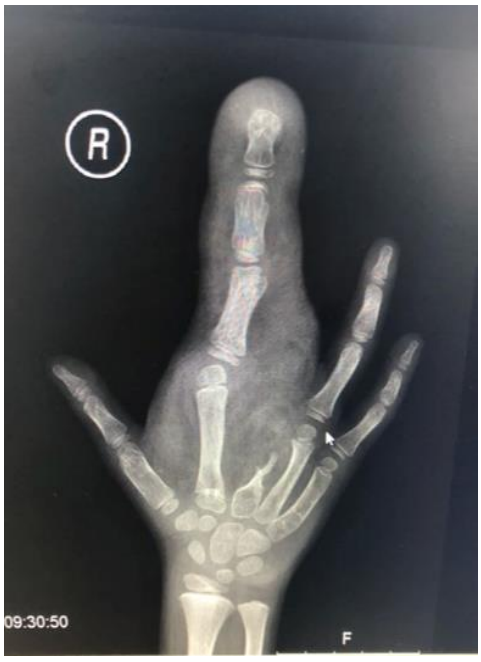

Figure 7 Bony hypertrophy resulting in focal gigantism

MRI revealed overgrowth of the bones and fatty tissues of the index finger (Fig. 8). Hypertrophic degenerative changes were seen in the interphalangeal joints with osteophytes and joint space loss. There was no vascular malformation or focal soft tissue mass. The ideal principle remains undisclosed yet, but the most likely mechanism presumed to be is congenital exaggeration and overgrowth of fibroadipose tissue of the perineurium layer. Lipomatous degeneration and fetal vasculature abnormality are other hypotheses that have been suggested. The correlation between fibrolipomatous hamartoma and macrodystrophia lipomatosais vague and ambigous; the similar MRI hallmarks and histological appearance of fat proliferation and nerve fibrosis make it complicated to differentiate the two conditions. Between 22% and 67% of patients with fibrolipomatous hamartoma may have also inherited macrodystrophia lipomatosa at birth. The differential diagnosis includes neurofibromatosis type I, lymphangiomatosis, haemangiomatosis, Klippel-Trenaunay-Weber syndrome and proteus syndrome. The clinical history and a thorough physical examination can help differentiate these conditions.

The aims of principal management of fibrolipomatous hamartoma with macrodystrophia lipomatosa are symptomatic relief and cosmetic considerations. In case of amputation, the affected nerve could be obliterated. Several debulking surgeries are often required (Brodwater et al., 2000). Surgical biopsy is redundant for fibrolipomatous hamartoma lesions that divulge the classical MRI hallmarks that resemble a coaxial cable on the axial view and spaghetti on a coronal or sagittal view (Tahiri et al., 2013). A regular follow up is a must as the recurrence rate is about 33% to 60%.

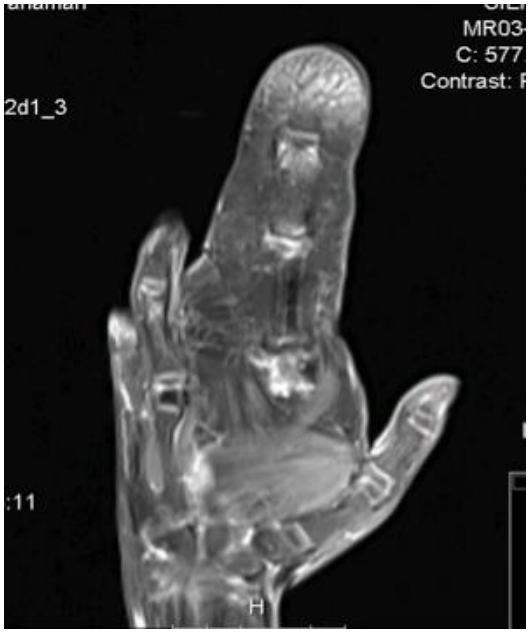

Figure 8 Overgrowth of the bones and fatty tissues of the index finger

4. CONCLUSION

Finally, fibrolipomatous hamartoma with macrodystrophia lipomatosa is an infrequent inborn condition. These conditions co-exist in 40% of cases. It can involve any peripheral nerves but most commonly affects the plantar and median nerves. Radiographic studies are sufficient for accurate diagnosis and allow avoidance of unnecessary biopsy. Amputation of the affected finger for functional and cosmetic purposes is the main goal of treatment.

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Author Contributions

Awan B, Aljaaly H performed the surgery and follow-up of the patient.
Altumaihi F, Altumaihi R, Oufi R reviewed the concept and collected data.
All authors participated in the writing and revision of the text.

Informed consent

Written & Oral informed consent was obtained.

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Conflict of Interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are presented in the paper.

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