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Case report

Surgical Management of Macrodystrophia Lipomatosa, a rare case report of the left hand middle finger macrodactyly

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ABSTRACT

Introduction: Macrodystrophia Lipomatosa is a rare congenital non-hereditary disorder, characterized by the presence of gigantism of a small part of the limb or the whole limb due to an overgrowth and disproportionate growth of fibroadipose tissue, causing macrodactyly.

Case presentation: One case of the 14-year-old girl presented with an enlarged middle finger of her left hand since birth, discomfort during the last 6 months, and resistance to flex, was underwent surgery by performing debulking procedure and a yellow cord-like mass was obtained from the digital nerve covered and enlarged by fibrofatty tissue to the palm area, could be cut off completely. Imaging and histopathological examination revealed to macrodystrophia lipomatosa. We follow up the patient for the finger's range of motion, the neurological disturbance and re-enlargement of the tumor.

Discussion: Since there were compression of the nerves, functional impairment due to enlarge fingers and cosmesis problems, the surgery was indicated. Types of surgery may include debulking of soft tissue, especially adipose tissue. The mass size was decrease, motor function and movement of the middle finger were normal post operatively, but sensory deficits persisted according to the distribution of the digital nerves. Three months and six years after surgery, no enlargement of the middle finger, normal motor function, normal finger movements but sensory deficit still persisted according to the distribution of the digital nerves.

Conclusion: Surgical management in macrodactyly due to macrodystrophia lipomatosa of the finger give the satisfactory result.

1. Introduction

Macrodystrophia lipomatosa is a rare, non-hereditary congenital disorder. Characterized by the gigantism of a small part of the limb or the whole limb due to an overgrowth and disproportionate growth of fibroadipose tissue [1,2]. This disorder can affect pediatrics patients, adults and elderly, and a congenital disorders affecting the upper and lower extremity [3,4].

In pediatric patients several terms are often used for this condition such as macrodactyly, megalodactyly, digital gigantism, macromelia, partial acromegaly, and macrosomy. Macrodactyly is a term used to described a state of disproportionate enlargement of the finger from birth or starting in the first years of life. Both soft tissue and bone are thoroughly enlarged in the affected finger [5,6].

Surgical management can be performed for this case to remove the mass that make some problems in the affected finger. We present a rare case of the left hand middle finger macrodactyly due to macrodystrophia lipomatosa by debulking surgery management. The surgery was performed by hand surgeon with 3 years experiences. Significant improvement of range of motion, neurological disturbance and no enlargement of existing tumor were used to assess outcome of the surgery. The SCARE criteria is used for reporting this case report [7].

2. Case presentation

A 14-year-old girl came to the outpatient clinic complaining of discomfort on the tip of the middle finger of her left hand since 6 months ago. Initially the patient complained her middle finger appeared larger

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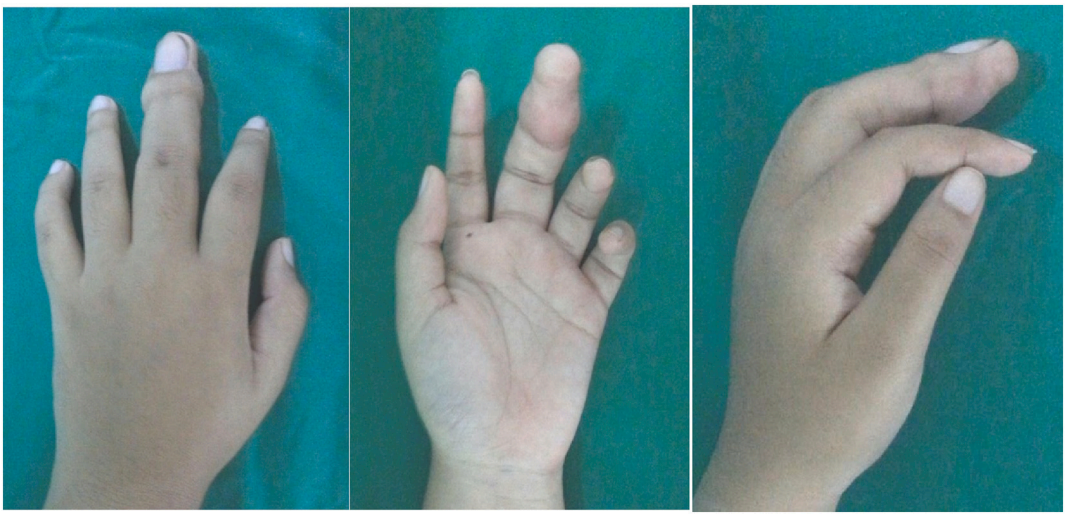

Fig. 1. Clinical appearance of affected hand.

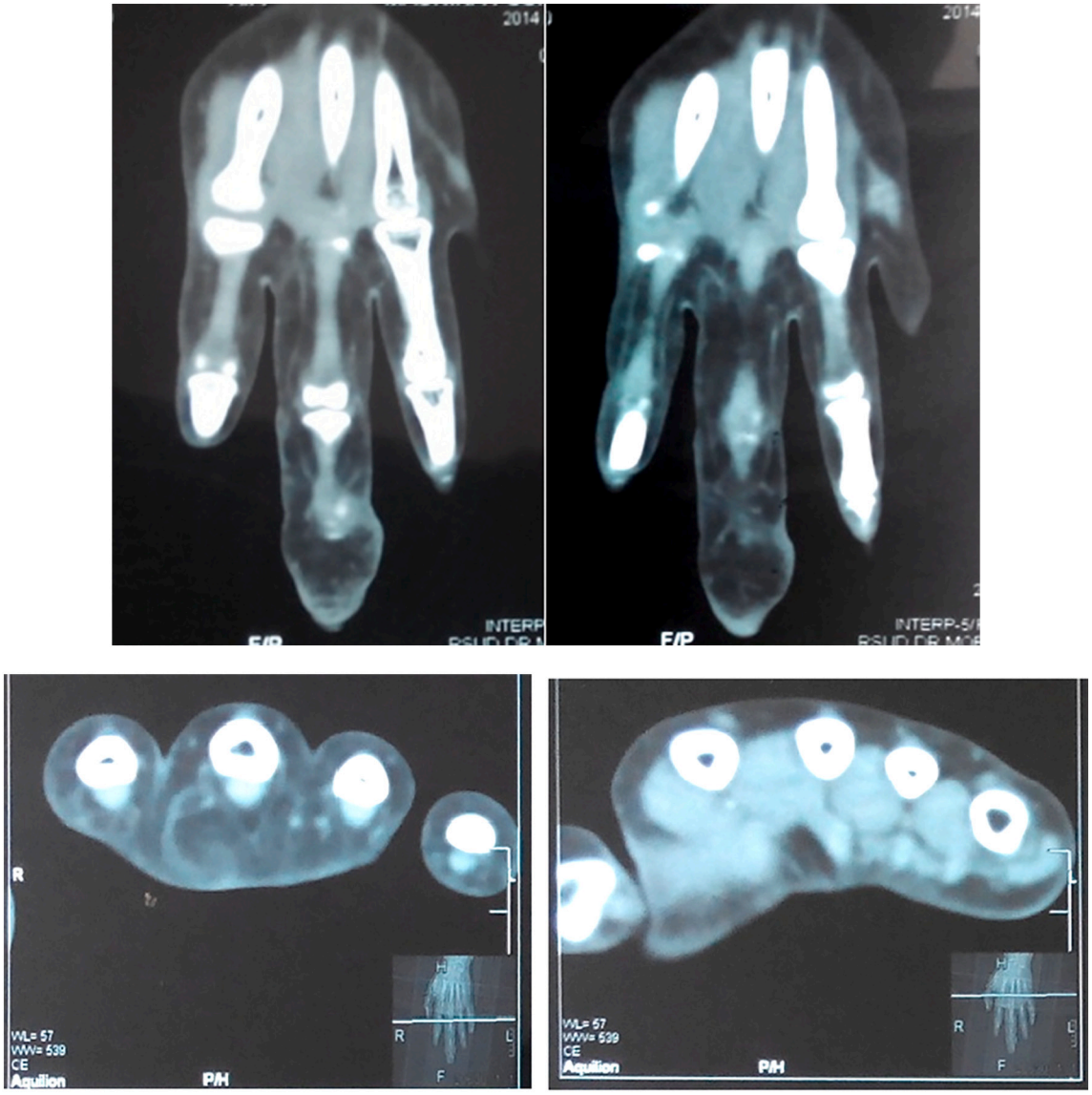

Fig. 2. CT-scan.

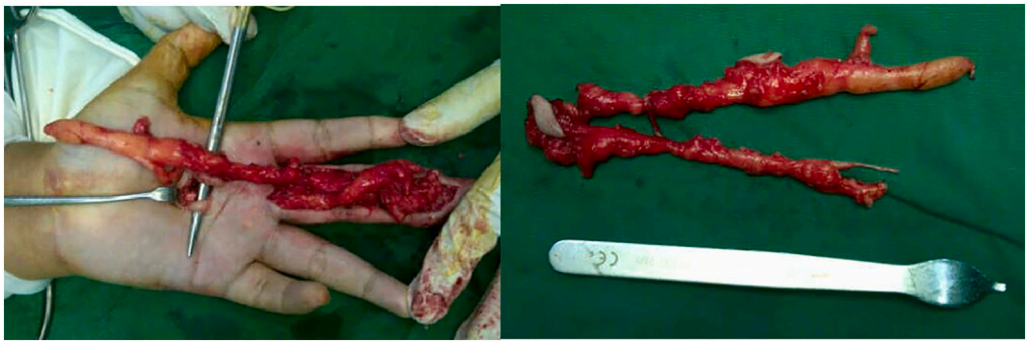

Fig. 3. Intra operative.

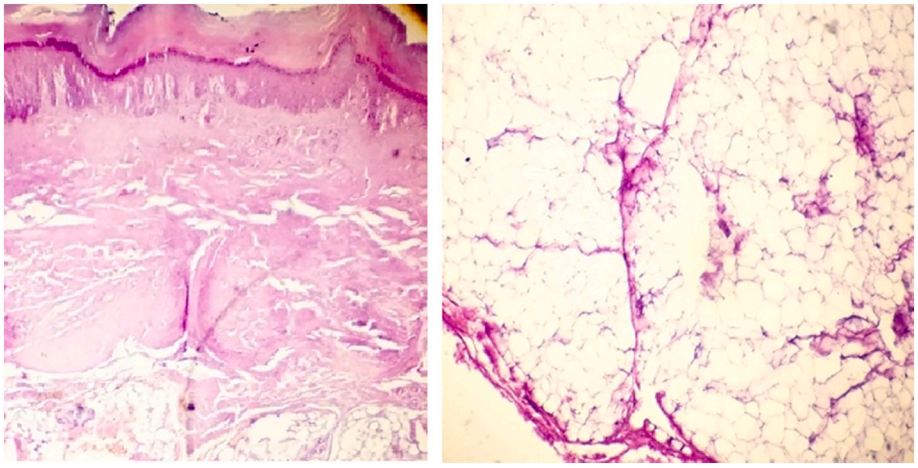

Fig. 4. Histopathology finding.

than other fingers from birth and grew larger with age. Since 3 months ago, she started complaining of tingling sensation at the fingertip, and also limited motion of the finger joint. No history of trauma, no fever, no weight loss, no change of appetite, and activity daily living as usual.

Physical examination generally revealed no abnormalities. Examination of the local status of the left-hand region revealed an enlarged middle finger, the same color as the surrounding skin, no venectation was found. There was tenderness in the entire enlargement area, the temperature was the same as the surrounding skin, and hypo aesthesia in the distal part. Resistance to joint motion during flexion in the proximal interphalang, and distal interphalang joints was found, so the middle finger cannot flex fully (Fig. 1).

CT-scan showed an overgrowth of the middle finger involving the proximal, medial, and distal phalanges along with the surrounding soft tissue, with trabeculation structures of the bone that is still normal and there is no abnormal vascular image suggesting a localized gigantism involving the bone structure and surrounding tissue due to the presence of lipomatous macrodystrofia (Fig. 2).

Surgery had performed under general anaesthesia and used tourniquet, zig-zag incision at the volar site started from distal to proximal phalang. A yellowish fat-like tissue is found that forms a tube-like structure extending to the palmar region, then the incision was extended to the proximal palmar region. Debulking of the entire tissue was carried out, preserving the surrounding nerves and blood vessels. The tissue was taken for a histopathological examination. After surgery, the patient was given antibiotics and analgesics, periodic wound care, and joint exercises were performed, especially extension flexion on the middle finger. The mass size was decrease, motor function and movement of the middle finger were normal post operatively, but sensory deficits persisted according to the distribution of the digital nerves

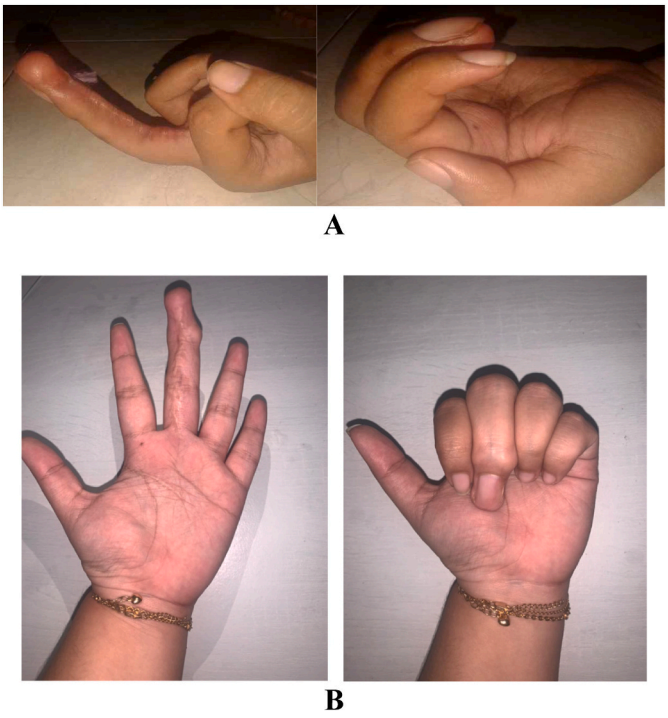

Fig. 5. Clinical appearance in the three months (A), and six years (B) after surgery.

(Fig. 3).

From the histopathological examination, there was a fatty tissue tumor which was limited by a septa-like connective tissue capsule, and there were several fibrotic blood vessels and fibrous connective tissue between the fatty tissue suggesting a lipofibroma (Fig. 4).

Three months after surgery, the surgical wound was good in condition, there was no enlargement of the existing tumor, and no resistance to joint motion on the middle finger. Six years later, no recurrency found in the middle finger and was able to fully flex. There were improvement range of motion in three months and six years after surgery comparing to the pre-operative condition. But sensory deficit still persisted according to the distribution of the digital nerves (Fig. 5).

3. Discussion

Macrodystrophia lipomatosa is a non-hereditary congenital disorder affecting a small part of the limb/finger or the entire limb which is seen as localized gigantism. Characterized by a complete enlargement of the fingers, usually the thumb and index finger, caused by an abnormal proliferation of fibroadipous tissue, fatty infiltration and hypertrophy of all components of the finger, including skin, bone, and nerves [8].

Clinically, this lesion is present at birth and is often recognized in the first 3 years of life. There is enlargement of the fingers or extremities that tend to be fast (progressive type) or there is consistent enlargement (static type), causing cosmetic abnormalities. This disorder is generally unilateral but can be more than one finger. The index finger is the most frequently affected finger, and may be accompanied by enlargement of the thumb, middle or ring finger. When it occurs on the thumb there is usually a radial deviation from the present deformity and is characterized by abduction and extension. If it occurs on 2 fingers it is usually a divergent deviation. There is bone enlargement and this deviation usually continues until the epiphyseal plate closes. The soft tissue will continue getting bigger with age [3,5,8].

There are complaints related to neurovascular compression due to swelling and may be accompanied by pain or not, degenerative changes, and impaired/loss of function of fingers or limbs. The presence of this enlargement will also tend to make the joint relatively stiffer, thus limiting the movement function of the affected limb [3,8]. Some cases may be accompanied by the presence of syndactyly, clinodactyly, brachidactyly and polydactyly [5,6].

Pathologically, macrodystrophia lipomatosa is characterized by an increase in all mesenchymal elements which are dominated by adipose tissue accompanied by other tissues such as periosteum, bone marrow, nerve sheath, muscle, and subcutaneous tissue [1].

Microscopic examination revealed an increase in the amount of subcutaneous fat, enlargement and excessive growth of nerve cells [2,5,9].

Radiological examination plays an important role in diagnosing this condition.

Plain radiograph shows soft tissue and bone hypertrophy with translucency in soft tissue due to increase adipose tissue. Distal phalanges are elongated and widened in the form of trapezoid or fungal-like formation with endosteal and periosteal bone deposition. Overgrown soft tissue is mainly found on the volar and distal aspects. The picture of secondary osteoarthrosis in the form of narrowing of the joint space, subchondral cysts and osteophytes are often found in adolescent or young adult patients [5,10].

Ultrasound examination shows tissue hypertrophy and the nerve involvement, hypertrophy distribution of adipose tissue (subcutaneous, intermuscular or intramuscular), and calcification, abnormal blood flow due to vascular malformations missed by plain radiographs [2,8].

A CT scan can be used to show fat proliferation along the course of the peripheral nerves. A CT scan will more clearly show excessive bone growth in the area supplied by the peripheral nerves [6,10].

MRI examination can show fibrous tissue and excess adipose tissue, hypertrophy of the bones and cortical thickening in the affected part of the body leading to exostosis of the affected bone [2,6,10].

Management can be done mostly by surgery. If the enlargement is not too serious, and there are no symptoms on the peripheral nerves, non surgery management by observation is made until the complete patient's growth [11].

Indications for surgical management are compression of the nerves, functional impairment due to enlarge fingers or extremities, and cosmesis problems. Variations in surgery are performed based on the patient's symptoms, age, and patient severity [12].

Types of surgery may include debulking of soft tissue, especially adipose tissue, followed by gradual reconstruction, compression of the peripheral nerve-releasing carpal tunnel, epiphyseodesis, reduction osteotomy or ray amputation. Debulking the tumor is not done extensively which will give bad results. The incidence of neurological injury after extensive debulking is 30% to 50% with a recurrence rate of 33% to 60%. The goals of epiphyseodesis and osteotomy are to stop abnormal growth of limbs and correct deformities due to disproportionate growth so as to get functional results. Attempts to maintain neurological function must be endeavored in the management of surgery. Amputation is performed when the affected limb cannot be maintained or the fingers cannot function properly after therapy, and to prevent further complications such as nerve entrapment in the future [2,6,8]. In this case we performed surgery by debulking to reduce the mass since there were a compression of the nerve indicated by tingling sensation at the fingertip, functional impairment due to enlargement of the finger by limited joint motion of the finger, and cosmesis problem.

The differential diagnosis of macrodystrophia lipomatosa in the presence of macrodactyly on the hands and feet is hemangiomatosis, Proteus syndrome, neurofibromatosis type 1 or neurofibromatosis plexiformis, lymphangiomatosis, fibrolipomatous hamartoma, Maffucci's syndrome, Ollier disease and Klippel-Trenauny-Weber syndrome. The suspicion of a benign and malignant tumor is due to the progressive enlargement of the soft tissue mass. Some benign tumors include lipomas, ganglion cysts, congenital syndromes (Dejerine Sottas and Weber syndromes) and traumatic neuroma. While tumors that are potentially malignant include schwannoma, neurofibroma and soft tissue sarcomas [13,14].

4. Conclusion

Surgical management in macrodactyly due to macrodystrophia lipomatosa of the finger give the satisfactory result. Postoperatively, the mass size decreased, no compression of the nerves, and no functional impairment due to enlarge fingers.

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The authors declare that this study had no funding resource. There is no sponsor for this study. All expenses are covered up by the author.

Ethical approval

Ethical approval has been issued by Orthopaedic Hospital Prof. Dr. R. Soeharso Surakarta Ethics Committee on April 26th, 2021 (No: LB.02.01/XXX..3/2690/2021).

Consent

Written informed consent obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

1. Name of the registry: Surgical Management of Macrodystrophia Lipomatosa, A Rare Case Report of the Left Hand Middle Finger

Macrodactyly

2. Unique identifying number or registration ID: researchregistry6942

Guarantor

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Data analysis and interpretation: Tito Sumarwoto, Mujaddid Idulhaq, Seti Aji Hadinoto, Asep Santoso, Dimitri R. Abimanyu.

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Declaration of competing interest

No potential conflict of interest relevant to this article was reported.