

PEDIATRIC MACRODACTYLY: A CASE REPORT

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INTRODUCTION

Macrodactyly is a rare congenital overgrowth deformity affecting the upper or lower extremity digits. Not only the fingers and toes can be affected, but also adjacent parts of the hand or foot [4]. It is more predominant in the region of the hands, especially in the second or third fingers. It often involves several types of tissues, such as: bone, nerve, skin, vessels and fat [1]. When present in the hand, it represents less than 1% of congenital anomalies in the upper limb. Two types of macrodactyly are described in the literature. Static macrodactyly, which is present from birth and whose fingers grow in line with normal fingers, and progressive macrodactyly, in which there is exacerbated bone growth at a faster rate than the other fingers [3]. It is assumed that the pathophysiology is due to genetic mutations in the AKT1, PIK3CA and PTEN genes, in addition to epigenetic factors [1]. In this sense, the disproportionate hypertrophy generated by macrodactyly, in addition to resulting in functional disability most of the time, can bring negative psychological harm to the patient, especially in a school environment [2].

The treatment for macrodactyly is surgical, with the aim of reducing symptoms and preserving the functionality of the limb. Surgical techniques are the most diverse and are selected based on the type, surgeon's experience and family expectations. Surgical options include: osteotomy or ostectomy; debulking-dissecting of bone and soft tissues; epiphysiodesis; nerve decompression or amputation [3]. Amputation, although frequently performed, carries a risk of skin necrosis, infection and recurrence, and must be avoided if possible [1]. Thus, the objective

of this study is to report the treatment of a case of macrodactyly in the left hand of a four-year-old child, with emphasis on the surgical technique adopted.

CASE PRESENTATION

Female patient, four years old, diagnosed with congenital macrodactyly in the third finger of her left hand (Figure 1).

There was no evidence of other congenital anomalies or other comorbidities. Patient born by natural birth, with normal, full-term pregnancy, without complications. For our patient, no genetic studies were performed to investigate macrodactyly.

On physical examination, a marked increase in the third finger of the left hand was observed, with a slight increase in the first and second fingers, in relation to the contralateral hand. The third finger showed flexion and extension movements without limitation, as well as preserved sensitivity, in addition to showing medial bending. There was an increase in volume in the palm of the hand, in line with the 3rd finger. Vital signs and laboratory tests were within normal limits, with no changes.



Figure 1: Macrodactyly of the third finger of the left hand.

The surgery was based on reconstruction of the 3rd finger, with reinsertion of the flexor tendon, arthrodesis and debulking-dissecting, under general anesthesia. The 3rd finger was reconstructed by carrying out a careful dissection of the tissues, always paying attention to the identification of vital structures, such as the digital nerve and digital artery

We used a technique to shorten the finger, in which it was narrowed by degreasing and, after dissection, the lateral digital nerve was removed along with the skin that it innervates (Figure 2). Thus, the proximal part of the distal phalanx was removed and an arthrodesis of the distal interphalangeal joint was performed, with the resection of a portion of the middle phalanx (Figure 3). The nail bed that was initially removed was repositioned at the end of the operation and sutures were placed to bring the finger closer together, which allowed normal alignment of the 3rd finger, without the need for amputation.



Figure 2: Identification of the digital nerve.



Figure 3: Arthrodesis of the distal and middle phalanx was performed, with the deep flexor tendon disinserted.



Figure 5: Result after 2 months.

In the immediate postoperative period, a reduction of up to 25% of the finger was possible, with a reduction in ulnar deviation and a pertinent aesthetic appearance, as it presents symmetry with the contralateral hand (Figure 4).

Follow-up after 2 months revealed a functional finger, that is, with active movement and preserved sensitivity, with satisfactory healing characteristics, without signs of infection, dehiscence or other postoperative complications (Figure 5).



Figure 4: Immediate post-operative period.

DISCUSSION

The diagnosis of macrodactyly is clinical and involves the identification of the phenotypic characteristics specific to the condition.

The patient in question presents a marked increase in the third finger of the left hand, with a slight increase in the first and second fingers. The patient had no positive family history for the syndrome, being the first in his family to present the condition.

The surgical procedure was performed in a single surgical procedure, respecting the patient's physical limits and his young age. With the aim of reducing the volume of the third finger and preserving extension and flexion movements, nail growth and sensitivity. The technique used to perform the surgical procedure is well established in the literature, with the modified Hoshi technique being used, as, in this procedure, there was no need for metacarpal osteotomy as described in the original technique, thus, it was possible to reduce phalanx deviation only with realignment in arthrodesis of the distal and middle phalanx, in addition to a good dissection of the digital nerve that was stimulating tissue growth.

This way, the treatment became possible and, when discussed with the orthopedics team, they advised the amputation of the third finger as the only alternative.

CONCLUSION

This case report highlights the complexity of treating macrodactyly in pediatric patients and the importance of a personalized approach. The surgery performed on the patient, using a modified Hoshi technique, with arthrodesis and degreasing, proved to be effective in reducing the volume of the affected finger and preserving its functionality, without the need for amputation. The success

of the procedure highlights the importance of a careful evaluation and the choice of appropriate surgical techniques, always aiming for the best functional and aesthetic prognosis for the patient.

Continuing follow-up is essential to monitor progress and prevent possible complications. This case reinforces the need for individualization of treatment and the potential for less invasive techniques to achieve optimal results in similar cases.

DISCLOSURE

The authors report no conflicts of interest in this work.

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