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Case Report

Hand Intrinsic Hemihypertrophy Associated with Absence of Carpal Tunnel and Hand Anomalies - Case Report

Paulo Henrique Paladini Filho MD[1] and Luis Guilherme Rosifini Alves Rezende MD, PhD[2]*

[1]*Hand Surgeon and Associated Research of the Hand Surgery, Microsurgery and Upper Limb Reconstruction Division of Ribeirao Preto Medical School of the University of Sao Paulo, Brazil*

[2]*Professor and Head of the Hand Surgery, Microsurgery and Upper Limb Reconstruction Division and Fellowship Program of Ribeirao Preto Medical School of the University of Sao Paulo, Brazil*

***Corresponding author:** Luis Guilherme Rosifini Alves Rezende, Department of Orthopedics and Anesthesiology, Ribeirão Preto Medical School of the University of São Paulo. Ribeirão Preto, São Paulo, Brazil

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Abstract

Hand hemihypertrophy is a rare condition when the intrinsic muscles are affected. A few cases explain the management. We present a case report of a three-year-old boy who had intrinsic hand hemihypertrophy with anomalies in the carpal and Guyon channels and loss of hand function. The selective operative soft tissue debulking was performed, with hand improvement and recovery of grasp function.

Keywords: Macrodactyly; hemihypertrophy; carpal tunnel; median nerve

Introduction

Macrodactyly is a rare congenital condition defined by the abnormal overgrowth of one or more orthopedic fingers. Both soft tissue and bone are diffusely enlarged. Its most common manifestation is an isolated anomaly associated with lipofibromatosis of the nerve [1-4]. Gigantism can be classified as segmental, in which only part of a limb is affected, or hemihypertrophy, with one side of the body affected. The latter may be subtle, and if there is macrodactyly of all the fingers on the corresponding side, the remainder of the limb should be carefully examined for enlargement. This study aims to report a patient with macrodactyly associated with changes in the hypothenar region of the palm and the absence of carpal tunnel [5-10]. Few studies explain hand hypertrophy, keeping this a rare and unique differential diagnosis with few cases described.

Case Report

A 3-year-old boy whose parents sought a pediatrician due to a deformity in his right upper limb since birth. He presented hand muscle hypertrophy throughout the limb, from the pectoral muscles to the palm, with overlapping of the 2nd and 3rd fingers. Parents complained of difficulty holding objects due to hypertrophy of the hypothenar portion of the right hand, requiring the use of the non-dominant (left) hand for daily activities. The pediatrician did not find any other changes in height and expected weight for his age. The physical examination revealed hemihypertrophy of

the entire right upper limb with flexion of the 3rd and 2nd fingers, with overlapping of the second finger over the third finger. The hypertrophy of the hypothenar area of the hand obstructed the grasp and pinch (Figure 1). There were no functional changes in the shoulder and elbow. There were no changes to sensitivity.

The MRI of the right upper limb showed no changes in bone, muscle, neurovascular and subcutaneous tissue (Figure 1). Due to the difficulty in using the hand and the school age, we opted for soft tissue debulking procedures of the hypothenar region. The approach to the hypothenar region showed intense hyperplasia of the hypothenar muscles (palmaris brevis), which was resected (0.128kg), allowing us to confirm the absence of the Guyon canal, as well as the absence of the transverse carpal ligament, where the median nerve could be visualized. Furthermore, the myotendinous junctions of the flexors and intrinsic muscles were hypertrophic and challenging to distinguish. The median and ulnar nerves were enlarged, with thicknesses similar to the average adult size. A persistent median artery and an aberrant recurrent branch of the median nerve also presented anomalies. The release of the second web incompletely syndactyly with repositioning and correction of the overlap of the index finger was performed (Figure 2). The patient had a three-year follow-up with no recurrence and good function outcomes improving his function (Figure 2).

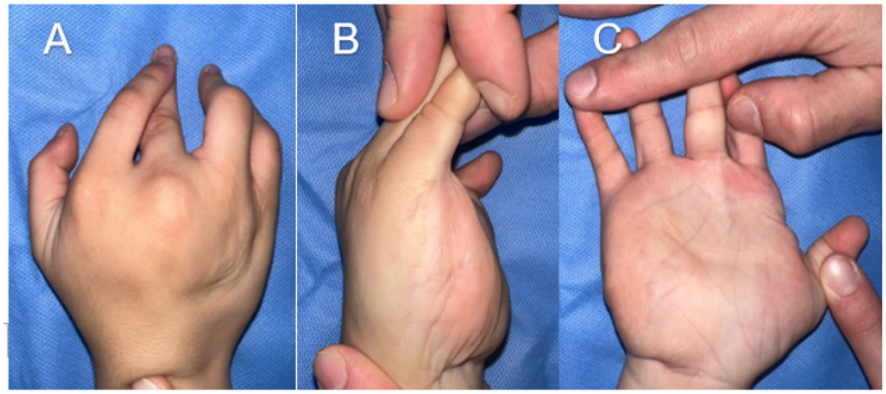

Figure 1: Hand hemihypertrophy of the hypothenar eminence, with simple incomplete syndactyly of the second web. A. Volar view (hypothenar hypertrophy), B. Dorsal view and C. Ulnar view. Courtesy of Professor Luis Guilherme R. A. Rezende Ribeirão Preto Medical School at the University of São Paulo.

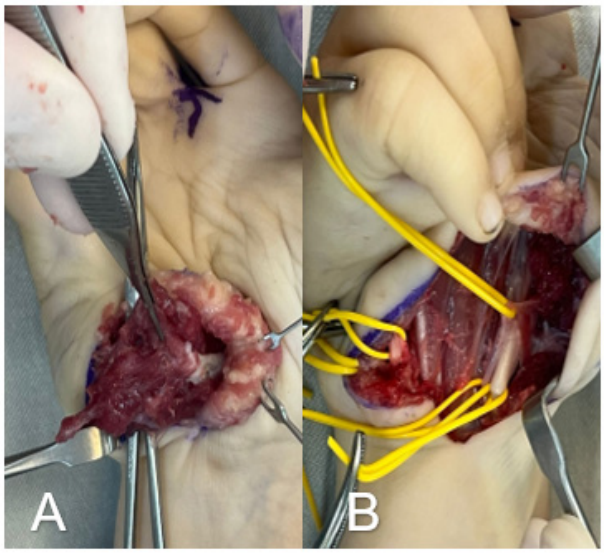

Figure 2: Soft tissue debulking procedure. A. Volar view (hypothenar hypertrophy), B. Aberrant and absent Guyon Channel and Transverse Ligament of the Carpal Tunnel, with the median nerve without carpal tunnel with hyperplasia of the finger's flexor tendons, Ribeirão Preto Medical School at the University of São Paulo.

Discussion

Macrodactyly is a rare condition characterized by abnormal growth of one or more fingers or body parts. Although it is an uncommon condition (0.9%), it reduces patients' function and quality of life. Its etiology is unknown; however, overgrowth is believed to be related to abnormal nerve supply. The lipofibromatosis is the most common subtype where fatty infiltration and enlargement of the nerve and finger are observed. It can be static or progressive. Static is present at birth, with proportional growth of the fingers. Progressive is the most common type involving changes in one or more fingers, with angular deviation. However, it doesn't explain the usual finger length in our patient. Hand hemihypertrophy is a scarce condition related to the hand's intrinsic muscles, being hard to classify in the Flatt classification [11]. The modified Flatt Classification has four subtypes. However, its management is challenging and selective soft tissue debulking is necessary to improve hand function. Type IV is appropriate to describe our case. The management must consider the expectations of the parents and, sometimes, the child. The possibility of recurrence and poor hand function must be addressed. Among the surgical options chosen for this patient due to hypertrophy of the hypothenar region, the soft tissue debulking procedure was an alternative that promoted improvement in function without recurrence after two years of postoperative follow-up. However, the anomalies observed in this patient make the need for further studies on this condition peculiar [12].

Conclusion

We conclude that one of the macrodactyly presentations is the intrinsic hand muscles hemihypertrophy without the typical involvement of the fingers.

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Hospital of Clinics of Ribeirão Preto Medical School of the University of São Paulo, Brazil.

Conflict of Interest

No conflict of Interest.

References

1. Scott W Wolfe, William C Pederson, Scott H. Kozin, Mark S Cohen (2021) Green's Operative Hand Surgery. 8th Edition, Philadelphia, Elsevier. Part IV Chapter 36 Deformities of the Hand and Fingers Pp. 1438-1448.
2. Batista KT, Cauhi AF, Almeida CEF, Araújo EA, Jesus GÂB, et al. (2008) Hand macrodactyly: a case report. Rev Bras Cir 23(2): 128-130.
3. Kotwal PP, Farooque M Macrodactyly (1998) Macrodactyly. J Bone Joint Surg Br 80(4): 651-653.
4. Joshua S Gluck, Marybeth Ezaki (2015) Surgical Treatment of Macrodactyly. J Hand Surg Am 40(7): 1461-1468.
5. F Fitoussi, B Ilharreborde, P Jehanno, J M Frajman, P Souchet, et al. (2009) Macrodactyly. Chir Main 28(3): 129-137.
6. AJ Barsky (1967) Macrodactyly. J Bone Joint Surg Am 49(7): 1255-1266.
7. Marybeth Ezaki (2019) Insights into the pathogenesis of macrodactyly. J Hand Surg Eur Vol 44(1): 25-31.
8. JA Herring, VT Tolo (1984) Macrodactyly. J Pediatr Orthop 4(4): 503-506.
9. V Kalen, DS Burwell, GE Omer (1988) Macrodactyly of the hands and feet. J Pediatr Orthop 8(3): 311-315.
10. João Nunes da Costa, Júlio Matias (2015) Segmental Hemihyperplasia-Related Macrodactyly with Congenital Renal Agenesis: A Hand Surgeon's Point of View. Arch Plast Surg 42(5): 655-658.
11. Flatt AE (1994) Large fingers. Flatt AE (eds.)., The care of congenital hand anomalies, 2nd edition. St. Louis, Quality Medical Publishing.
12. Semih Takka, Kazuteru Doi, Yasunori Hattori, Izuru Kitajima, Kazufumi Sano (2005) Proposal of new category for congenital unilateral upper limb muscular hypertrophy. Ann Plast Surg 54(1): 97-102.