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MACRODACTYLY: REPORT OF EIGHT CASES OF A RARE ANOMALY*

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SUMMARY: Kostakoğlu N, Kayıkçıoğlu A, Şafak T, Özcan G, Keçik A, Gürsu G. (Department of Plastic and Reconstructive Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey). Macrodactyly: report of eight cases of a rare anomaly. Turk J Pediatr 1996; 38: 73-79.

Macrodactyly or megalodactyly is a rare anomaly of the extremities. Neural factors are involved in the etiology. Presented here are eight cases which comprise five macrodactylous toes and three fingers. The mean age at first referral was 6.8 years. Six patients underwent resection of the proximal phalanges together with bulk reduction of the soft tissue mass. Only soft tissue reduction was performed in the remaining two patients. Skin necrosis was observed in two cases, one of which necessitated amputation at the proximal interphalangeal joint level. The functional outcome was evaluated as poor with limited range of motion and stiffness in the joints. As far as functional results are concerned, macrodactylous toes had a better prognosis than that of fingers.

It was concluded that none of the available methods as yet gives ideal functional and cosmetic results in macrodactyly. *Key words: macrodactyly, megalodactyly.*

Macrodactyly is overgrowth of the toes and fingers. Local gigantism, megalodactylia, megalodactilism, digital gigantism, macrodystrophia lipomatosa and localized hypertrophy have been used as synonyms for macrodactyly[1-5]. It is the least common congenital anomaly of the extremities, comprising 0.9 percent of the total number of reported cases in the literature[4, 5].

In many cases macrodactyly may be noted at the time of birth, whereas in some others abnormal growth appears soon after birth and often reaches gross proportions and constitutes a severe physical and psychological handicap. It is not always symmetrical. The finger or toe may tend to deviate toward the healthy side in cases where only one side of the finger becomes excessively hypertrophic. The etiology remains unexplained, with males affected more often, and association with other anomalies and family history is rare[1-5].

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In true macrodactyly, all components of a digit are enlarged. It must be distinguished from other pathologies resulting in enlarged digits such as hemangiomas, arteriovenous malformations, congenital lymphedema and lipomas (Table I)[4, 5].

Table I: Other Pathologies Resulting in Enlarged Digits

Hemangiomas
Arteriovenous malformations
Congenital lymphedema
Lipomas
Multiple enchondromatosis with hemangiomas
Klippel-Trenaunay-Weber syndrome
Osteoid osteoma
Melorheostosis

In this article, eight cases of macrodactyly are presented and a comparison of results is made by reviewing the literature.

Material and Methods

Between 1965 and 1994, eight cases of macrodactyly were treated in our clinic. During this period, a total of 650 extremity anomalies were examined. In our series, macrodactyly consisted of 1.23 percent of these cases.

The age of the patients ranged from one to 25 years; five were female and three male. The patients all described the pathology as being present since birth, and all were operated on after their first referral to our clinic. The mean age at operation was 6.8 years.

Results

The patients are listed in Table II. In five of the patients the foot was the site of the anomaly, while in the remaining three the hand was affected.

Table II: List of Patients

Age of First Operation	Sex	Site	Digit	Assoc. Anomaly	Family History	Consanguinity
5	F	RF	2-3	–	–	–
3	F	LF	2	–	–	–
25	M	LF	4-5	–	–	–
2.5	F	LF	2-3	–	–	–
13	F	RH	3	–	–	–
1	M	LH	4	–	–	+
1	M	RH	3-4	+	–	+
4	F	RF	1-2	–	–	–

RF: right foot. LF: left foot.
RH: right hand. LH: left hand.

There was no positive family history in any of the patients. However, in two cases the parents were first and second degree relatives.

In one case, incomplete syndactyly of the long and ring fingers accompanied the macrodactyly (Figs. 1-4). Five out of eight patients had two digits involved, whereas in the remaining three only one digit was involved. The second and third rays were the most commonly affected digits.

The digital nerves were wound to be hypertrophic in all cases, one of which had hypertrophy of the soft tissues extending proximally to the palmar aspect of the hand; on exploration this was found to be due to hypertrophy of the median nerve starting from the entrance of the carpal tunnel (Fig. 5).

In five cases, partial resection of the proximal phalanges was performed together with reduction of soft tissue. Two cases underwent multiple reductions of soft tissue mass only. One had undergone amputation of the second ray of the foot by orthopedists at an early age, and underwent soft tissue resections in our clinic.

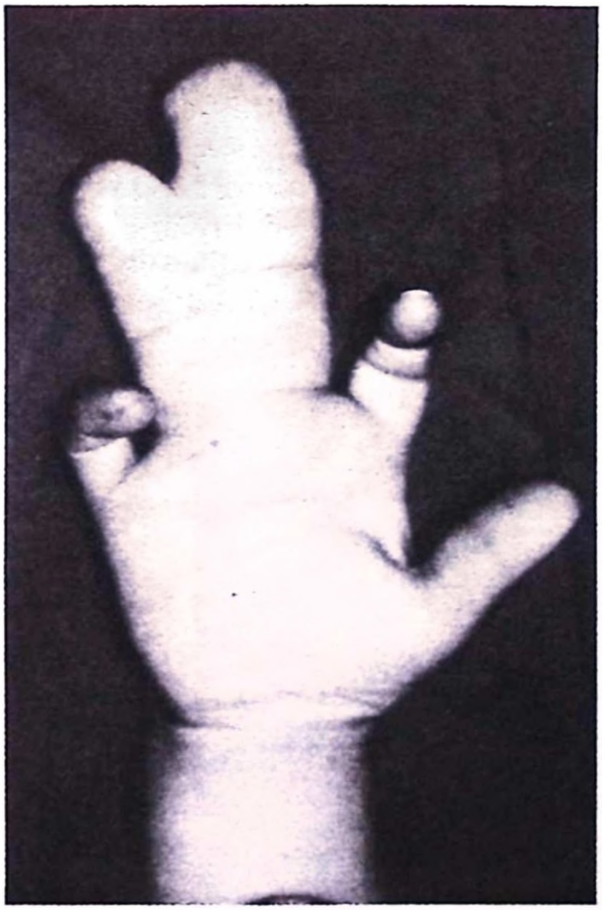

Fig. 1: Palmar view of third and fourth macrodactylous fingers together with incomplete syndactyly.

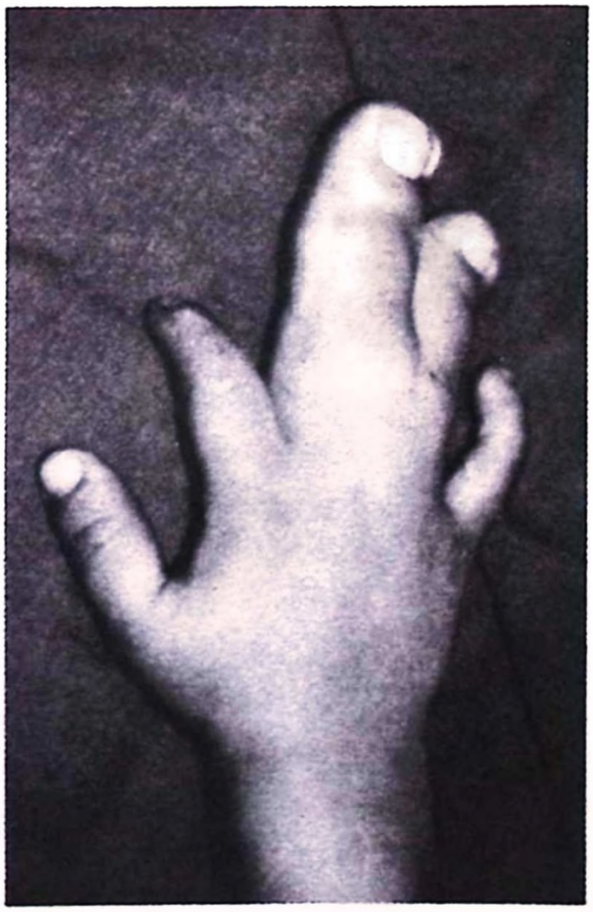

Fig. 2: Dorsal view of the same patient

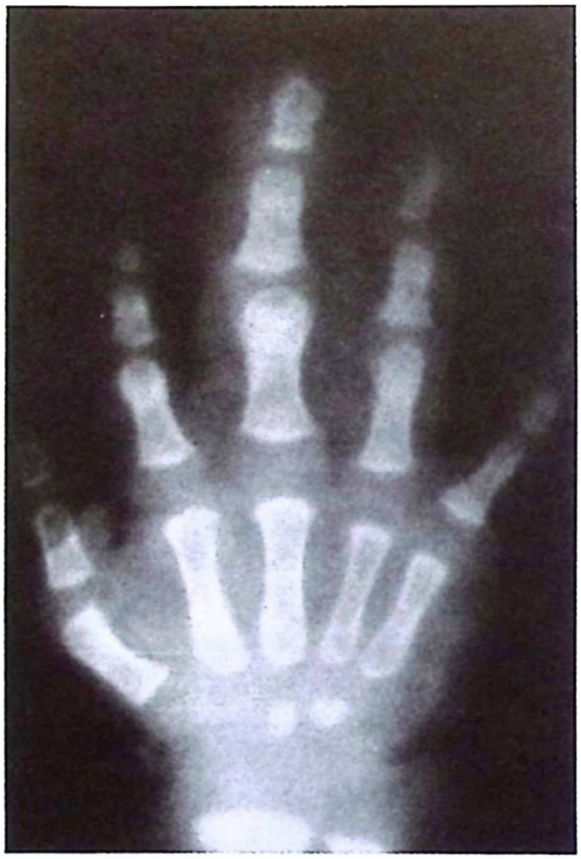

Fig. 3: X-ray of the macrodactylous hand.

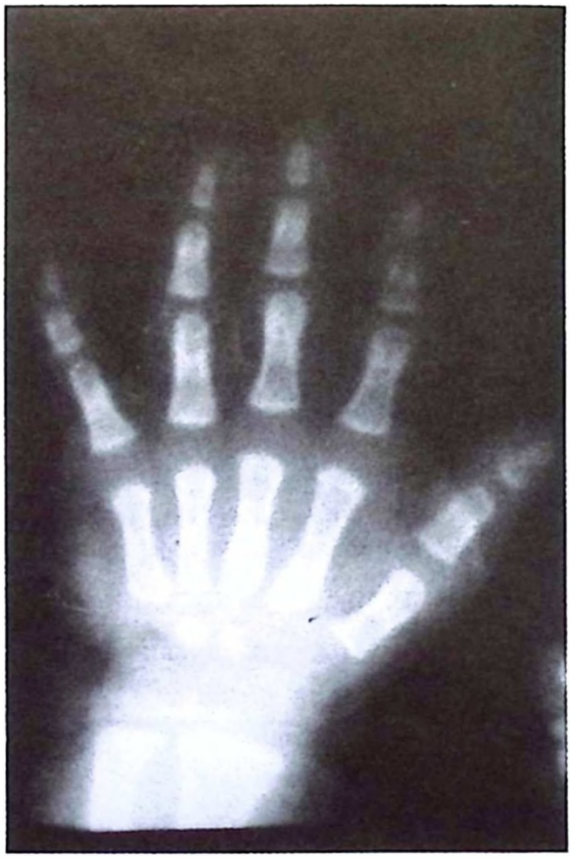

Fig. 4: X-ray of the normal hand of the same patient.

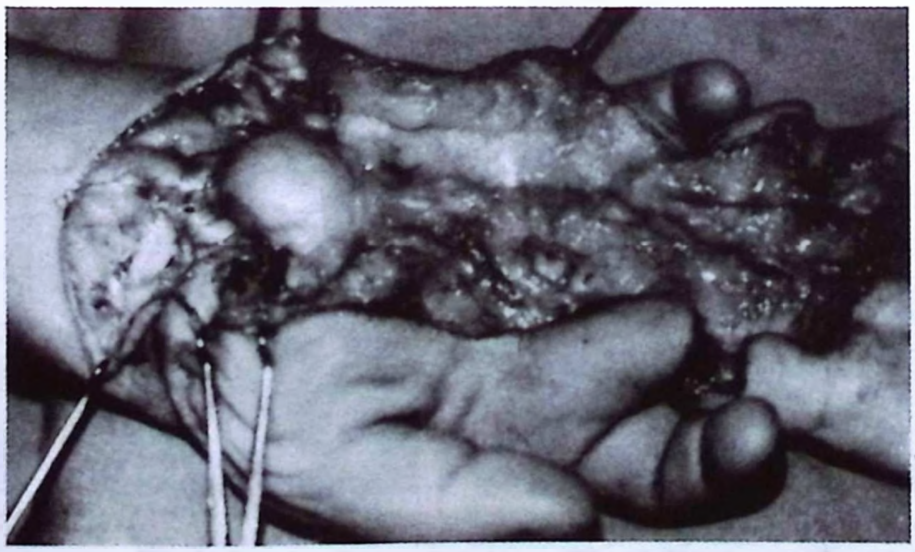

Fig. 5: Palmar soft tissue mass revealed to be a median nerve enlargement.

In two patients the postoperative course was complicated by necrosis of skin flaps. Supervening infection in one of these cases necessitated amputation of the long finger at the proximal interphalangeal (PIP) level. No further therapy was necessary in the other patient.

The functional outcome in macrodactylous fingers was poor, with limited range of motion and stiff joints which were of great concern in the upper extremity. In the foot this did not seem to be a problem since motion of the toes was rarely a matter of concern for the patient. Nearly all cases sought treatment mainly for cosmetic reasons; however, only two patients were satisfied with the final result, both of whom had macrodactylous toes. The remaining patients were neither satisfied nor wanted any more treatment.

Discussion

More than 300 fingers and 60 toes with macrodactyly have been reported[4]. Ninety-five percent of the cases were found to occur unilaterally[4]. No predilection for the right or left extremity was observed[3, 4]. The index finger was the most commonly affected finger, followed by the long finger, thumb, ring and little fingers in decreasing frequency. Involvement of multiple digits was found to occur more often than a single digit. Also, two digits were more frequently affected than three digits. The thumb and index finger, or the index and long finger, were the most commonly encountered combinations. Syndactyly was seen in 10 percent of cases[4].

In our series, macrodactyly of the toes was more common than the fingers. This is in contrast to the reports in the literature. A random refferral pattern of congenital anomalies in our country seems to be the responsible factor for this difference. We have also encountered no predilection for the right or left extremity.

The long and ring fingers and the second toe were the most commonly involved digits, and the finding that two digits were more commonly affected than three digits was also confirmed in our patients. The predominance of macrodactyly of the long and ring fingers in our series is in accordance with the report of Barsky[6]. However, Dobyns et al.[4] found the index finger to be the most commonly affected digit.

None of our patients had a family history of macrodactyly nor any other extremity anomalies. However, in two patients the parents were blood relatives. Macrodactyly is claimed not to be inherited, while association with other syndromes is reported to be a causative factor in inheritance as an autosomal dominant trait. Marriage between blood relatives is common in our country, and our two patients with consanguinity must have been incidental.

Treatment should be planned on an individual basis. Flatt[5] states that there is no correct age at which surgery should be started. Tsuge[1] advocates the inhibition

of finger growth when hypertrophy is not too marked, while shortening or reduction in size should be done after marked macrodactyly has developed.

In our series, the age at first referral ranged from one to 25 years. All patients but two were seen before five years of age. According to our experience, the disfigurement and malfunction caused by macrodactyly urges families to seek early intervention. Excluding the two late referrals, it seems that macrodactyly of the finger causes earlier awareness than macrodactyly of the toe.

A variety of operative strategies have been reported[1-6]. Dobyns et al.[4] recommends bulk reduction without any shortening of the bone for the young child. Tsuge[1] on the other hand reduces the length of the macrodactylous finger by excising the volar part of the distal phalanx and the dorsal portion of the middle phalanx, thereby the dorsal part of the distal phalanx and nail are recessed to fit the middle phalanx. If further shortening of the digit is required, he recommends excision of the MP joint.

When the size of the involved digit reaches the size of the same digit of the patient's parent of the same sex, epiphyseal arrest at the proximal end of each phalanx is advocated[5]. However, epiphysiodesis is reported to be extremely difficult and frustrating[4].

Our treatment policy was bulk reduction for the very early cases and addition of proximal phalangeal reduction in the older patients, which is a modification of the method for reduction of the thumb described by Millesi[7].

Tsuge[1] postulates that neural stimulation causes disproportionate growth of the finger. He therefore advocates stripping the branches of the digital nerves from the digital nerve trunk. The true incidence of median and ulnar nerve enlargement is unknown[1,4,8]. We have observed hypertrophy of the median nerve in one of our cases, which presented itself as a soft tissue mass in the palm. There is agreement in the literature that stiffening of the joint and degeneration of the bone results in a nearly motionless finger in most cases, and sometimes amputation is indicated for a huge digit that interferes with function of the rest of the hand[1,4,5].

Five of the macrodactylous digits in our series were located in the foot. Therefore, little concern was given to the functional outcome after surgery. In those cases with macrodactyly of the fingers, functional and aesthetic results were generally poor, and all patients required multiple operations. Skin necrosis was a serious problem in two cases, one of which necessitated amputation at the PIP level owing to supervening infection.

In conclusion, macrodactyly remains a challenge for the hand surgeon, and the perfect solution to the problem is yet to be found.

REFERENCES

1. Tsuge K. Treatment of macrodactyly. Plast Reconstr Surg 1967; 39: 590-599.
2. Thorne FL, Posch JL, Mladick RA. Megalodactyly. Plast Reconstr Surg 1968; 41: 232-239.
3. Jones KG. Megalodactylism. J Bone Joint Surg 1963; 45: 1704-1708.
4. Dobyns JH, Wood VE, Bayne LG. Congenital hand deformities. In: Green DP (ed). Operative Hand Surgery (3rd ed) Vol. 1. New York: Churchill Livingstone; 1993: 251-548.
5. Flatt AE. The Care of Congenital Hand Anomalies. St. Louis: Mosby; 1977: 249-262.
6. Barsky AJ. Macrodactyly. J Bone Joint Surg 1967; 49: 1255-1263.
7. Millesi H. A case study. In: Littler JW, Cramer LW, Smith JW (eds). Symposium on Reconstructive Hand Surgery, Vol. 9. St. Louis: Mosby; 1974: 173.
8. Allende BT. Macrodactyly with enlarged median nerve associated with carpal tunnel syndrome. Plast Reconstr Surg 1967; 39: 578-582.