

Clinical Study of Macroductyly

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=국문초록=

거대지의 임상연구

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정문상 · 박윤수 · 정필현

거대지 31례를 비교, 검토하였으며, 거대지는 특발성 및 속발성으로 구분되고 속발성에는 혈관종, 골연골중증등의 종양이나 그밖의 특이한 조직기형이 있다. 특발성의 경우 국소 비대외의 원인은 명확하지 않으며, 연부조직형, 골형 그리고 혼합형으로 분류될 수 있다. 거대지의 재건술에는 기능이나 외양이 중요하나 두 가지를 함께 유지하기는 힘들고, 반면에 기능이나 외양의 둘중의 하나를 유지하기는 훨씬 쉽다.

저자들의 방법에 의하면 적어도 한쌍의 혈관과 신경을 남기면서 수장부 지그재그나 편측성 피부절개술을 통하여 연부조직 감소술을 시행할 수 있었으며, 골에 대하여는 골단축술, 골단판제거술 그리고 유합술을 시행하였다. 저자들의 방법을 사용하여서는 합병증으로서의 절단이 없었으며 9례중 8례에서 거대지가 효과적으로 작아졌으며, 기능적인 면에서 Tsuge술식같은 복잡한 수술보다 좋은 결과를 얻었다.

Key Words: Macroductyly, Classification, Treatment

idiopathic type of macroductyly.

MATERIALS AND METHODS

Macroductyly is an uncommon congenital anomaly affecting the fingers and toes. The condition was first described by Power in 1840. Since that time over 109 cases have been reported.^{1,14,15)} In Korea 7 cases of macroductyly have been reported^{5,6,7,8,12)} No specific treatment has been established except amputation of part or all of involved digits before Bunnel suggested defatting procedure and epihyseal destruction in 1956. In 1967, Barsky¹⁾ has described two types of macroductyly: the static type in which the digit grows in proportion to the rest of body, and the progressive type in which the digit grows disproportionately to the rest of the body. Tsuge¹⁵⁾ has described a staged operating procedure for shortening of macroductyly in 1967.

The purpose of this report is to provide our classification and concept of treatment for the

* 본 논문은 1985년도 서울대학교병원 임상연구비 보조로 이루어진 것임.

We reviewed 31 patients with macroductyly, who were treated at department of orthopedic surgery, Seoul National University Hospital from 1975 to 1985. Male was affected in 14 and female in 17 cases. Average years of age at operation shows that operations were done at an earlier age group in idiopathic type. We could observe 20 cases of idiopathic macroductyly, and 11 secondary cases. Of the secondary type, hemangioma was most commonly associated (5 cases), followed by lymphangioma (2 cases), Ollier's disease (2 cases). Upon the clinical analysis of the 20 idiopathic cases, male and female were equally affected. The fingers were affected in 9 cases and toes in 11. There was no obvious difference between the right and left side, and in most of cases it showed unilateral involvement. Multiple digits involvement is predominant feature in our series. Index and middle finger were affected most often in hand, and second toe in foot. Follow-

Table 1. Clinical analysis of the idiopathic macrodactyly

Case	Age	Sex	Involved site	Type	Treatment	Complication	Follow-up (yrs + mo)	Result	
								Function	Appearance
1	6	F	*T 1,2,Rt.	***M	Amputation		10	###P	P
2	15	M	**F 2,3,Rt.	M	Tsuge op.	Amputation	8	P	P
3	17	F	F 1,2,Lt.	M	Tsuge op.	Amputation	8	P	P
4	8	M	T 2,3,4,Rt.	M	Amputation	+ + + IR	6	P	P
5	1	F	F 1,2,Rt.	M	Tsuge op.	IR	1	##F	P
6	10	M	T 1,2,Lt.	M	Tsuge op.	Amputation	6m	P	P
7	7	M	T 2,3,Rt.	M	Tsuge op.	Amputation	6m	P	P
8	7	F	F 2,3,Lt.	M	Our method	Clinodactyly	3	#G	G
9	5	F	F 2,3,Rt.	M	Our method		1	G	G
10	2	F	T 2,Lt.	M	Our method	Infection	3	F	F
11	1.3	M	T 1,2,Lt.	M	Our method		1	F	G
12	1.5	F	T 3,4,Lt.	M	Our method		1	F	F
13	0.3	M	T 2,3,Lt.	M	Our method	IR	6m	P	P
14	4	M	T 1,2,Lt.	M	Our method		3	G	G
15	4	M	T 2,Rt.	M	Our method		6m	G	G
16	22	F	F 1,Rt.	+ST	Excision		Lost	-	-
17	17	M	F 5,Rt.	ST	Excision		Lost	-	-
18	17	F	F 4,5,Rt.	ST	Excision		3	G	G
19	14	F	T 2,Both	+ + B	Metatarsal shortening		3	G	G
20	19	M	F 3,Rt.	B	No op.		-	-	-

* T: Toe, ** F: Finger, *** M: Mixed, +ST: Soft tissue, + + B: Bone, + + + IR: Ineffective reduction, #G: Good, ##F: Fair, ###P: Poor.

Fig. 1. Photograph (A) shows enlargement of the ring and little finger in the right hand, but X-ray (B) shows no bony enlargement.

up over 6 months was possible in 17 patients of idiopathic type and average duration of follow-up was 3.1 years (Table 1).

CLASSIFICATION

According to Barsky,¹⁾ true or idiopathic macrodactyly is a rare congenital malformation characterized by an increase in the size of all the element or structures of a digit or digits. In our series, there were 3 cases that had only enlarge-

Fig. 2. Medical photograph (A) shows elongated second toes in both feet without the enlargement of soft tissue, and x-ray (B) shows bony enlargement.

Fig. 3. Preoperative photograph (A) shows mixed type of macrodactyly in index and middle finger in the left hand. Three years after operation (B), the sizes of both fingers are markedly reduced. But there are some residual clinodactyly in the same fingers.

ment of the soft tissue without bony enlargement (Fig. 1), and there were 2 cases that had only bony enlargement without enlargement of the soft tissue (Fig. 2). We named this as bone and soft tissue subtype respectively. Fifteen cases that had the enlargement of all the elements were named as mixed subtype (Fig. 3). Macrodactyly that was associated with tumors and other specialized tissue malformations such as hemangioma, enchondromatosis, etc. might be classified as secondary type (Table 2).

TREATMENTS AND RESULTS

Until 1982, a variety of operations were used

Table 2. Classification of macrodactyly

Type	No. of patients
Idiopathic or true type	20
Soft tissue	3
Bone	2
Mixed	15
Secondary type associated with tumor	11
Hemangioma	5
Lymphangioma	2
Ollier's disease	2
Arterio-venous malformation	1
Maffucci syndrome	1
Total	31

Fig. 4. Preoperative x-ray (C) of the same patient, and x-ray at three years after operation (D).

Fig. 5. Initial photograph (A) shows secondary type of macrodactyly due to Ollier's disease. The bulky cartilagenous tissues were removed as completely as possible, and massive iliac bone grafting were performed. Three years after operation (B), the sizes of involved fingers are markedly reduced and the appearance and function of the hand have improved very much.

such as amputation, staged defatting of the soft tissue, epiphysectomy, and Tsuge's procedure etc. But there were too much complications such as necrosis of the digit, ineffective reduction and so on. In this situation a new method of treatment was highly needed. The operation must be simple to reduce the complication rate, and the aim of the

operation in this complex problem should also be simple. It seems better for a surgeon to choose either the preservation of the function or the improvement of the appearance as the aim of operation. In our series, all operations for both the appearance and the function resulted in failures such as amputations, ineffective reductions etc.

Fig. 6. Preoperative X-ray (C) shows enchondromatosis of both hands. Three years after operation (D) tumors are removed nearly completely and the PIP and DIP joint of left index and right middle finger are fused.

Out of 15 cases of mixed variety of macrodactyly, initial amputations were taken in 2 cases, Tsuge's operation in 5 cases. With Tsuge's operation necrosis of the involved digits were complicated and amputations were resulted in 4 cases. In one case the enlarged digit could not be reduced effectively. This means complete failure of the operation in our early series. In this dreadful situation we changed our policy of operation as follows:

Bulk reduction was tried with a volar zig-zag or unilateral single incision saving at least one pair of vessels and nerves. Shortening, epiphysectomy and fusion were done for bone (Fig. 3, 4). With this new method we could effectively reduce volume in 8 out of 9 cases, and there had been no amputations yet. In one case that the volume could not be effectively reduced, the digits were very huge. Even though we reduced about 60% of the length and 70% of

the circumference, the digits were still too huge. For 3 cases of soft tissue type, soft tissue excision was done. And for 2 cases of bone type, metacarpal shortening was done for one case that involved second toes bilaterally, and operation was refused by patient in the other case. As for secondary type of macrodactyly, excision of tumor and other appropriate operations were done case by case (Fig. 5, 6).

To evaluate the result of treatment, authors used our own criteria for the function (Table 3) and appearance (Table 4). And authors made scoring as follows: 2 point for Good, 1 point for Fair and 0 point for Poor. For 15 cases of the mixed idiopathic macrodactyly, Average scores are 0.2 for the function and zero for the appearance with Tsuge's operation. Average scores of our method are 1.4 for the function and 1.5 for the appearance.

DISCUSSION

The etiology of macrodactyly is unknown, but it has been postulated that impaired nerve function allows uncontrolled growth of other tissue.^{1,3,10,15)} Tsuge considered the hypertrophic adipose tissue and marked hypertrophy of the digital nerves as the most important finding in macrodactyly.¹⁵⁾ In hand, this condition most often involves fingers innervated by the median nerve.¹³⁾ So index and middle finger are affected most frequently and our series are also compatible.

Two types of macrodactyly have been described by Barsky.¹⁾ In the static type, an enlarged digit is present at birth and grows in proportion to the rest of the body. In the progressive type, the enlarged digit may not be present at birth but grows

Table 3. Criteria for the evaluation of the function

	Hand	Foot
Good	Above 2/3 of normal ROM	Little working problem
Fair	Between 1/3 and 2/3 of normal ROM	Walk one block without pain
Poor	Below 1/3 of normal ROM or amputated state	Walk one block with pain

Good (2), Fair (1), Poor (0)

Table 4. Criteria for the evaluation of the appearance

	Hand	Foot
Good	1. Effective reduction 2. Preservation of nail 3. Little gross deformity	1. wear same sized shoes 2. Preservation of nail
Fair	1. Effective reduction 2. Preservation of nail 3. Remained gross deformity	1. Wear different sized shoes but within one size difference 2. Preservation of nail
Poor	1. No effective reduction or amputated state 2. Absence of nail 3. Remained gross deformity	1. Wear different sized shoes over 2 size difference 2. Absence of nail 3. Amputated state

Good (2), Fair (1), Poor (0)

Table 5. Follow-up result, mixed idiopathic macrodactyly

	No. of cases	No. of amputation	No. of ineffective reduction	Function sum of score (average)	Appearance sum of score (average)
Amputation	2	2	0	0	0
Tsuge's op.	5	4	1	1 (0.2)	0
Our method	8	0	1	11 (1.4)	12 (1.5)

disproportionately to the rest of body. But Barsky's classification provide only a little benefit for the treatment. A new classification based upon treatment would be provided. In soft tissue subtype excision of the soft tissue is only required and in bone subtype a bone shortening is all that required. But in mixed subtype combined soft tissue and bone treatment is required.

There have been numerous reports of macrodactyly in whites^{1,2,9,14}) and a few report on blacks,⁹⁾ but little reports on yellows have been appeared in English literature. In most series the fingers are predominantly affected in whites, and the toes affected almost exclusively in blacks.⁹⁾ In our series, the fingers and the toes are affected almost equally (9 versus 11). There was no obvious difference of involvement between the right and left side, and in most of the cases it showed unilateral involvement. Krykman described that males are slightly more commonly affected than females and about 95 percent of the cases are unilateral.³⁾ Multiple digits involvement is two to three times more common as single digit involvement in both previous reports and our series. It is a uniform finding that the index and middle finger were affected most often in hand, and second toe in foot.^{1,4,14)}

According to our experiences, the most common and serious complication after Tsuge's or Barsky's procedure is the necrosis and subsequent amputation of the involved digit because of the bulk reduction by bilateral incision.¹⁵⁾ So it is highly recommended that incision must be unilateral or volar zig-zag which saves at least one pair of vessels and nerves. For bone, shortening, epiphysectomy and fusion can be done case by case.

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