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MACRODACTYLY-REPORT OF SIX CASES WITH REVIEW OF LITERATURE

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SUMMARY

Six cases of Macrodactyly between the age group of 4 and 14 years were investigated and managed surgically whereby excision of the excess tissue had been done. The affected children were having higher systolic blood pressure and invariably had excess of all the tissues, constituting the digit. The affected skin showed poor healing in 30% of the cases.

(Key Words : Macrodactyly, Angiography, A-V Communication.*)*

Macrodactyly ('large digit') is the term used for a disproportionately large digit apparent at birth or in early childhood. It is a rare congenital malformation characterized by an increase in size of all the elements or structures of a digit or digits. The phalanges, tendons, nerves, vessels, subcutaneous fat, finger nails and skin are all enlarged. Syndactyly is occasionally present. True macrodactyly should be distinguished from other forms of enlarged digits eg. hemangiomas, A-V malformations, congenital lymphedema, lipoma, Ollier's diseases, maffucci's syndrome, Klippel — Trenaunay, Weber syndrome, melorheostosis and osteoidosteoma.

Review of Literature

Klein described the first authenticated case of macrodactlyly in 1824. Flatt quotes an incidence of 0.9% in a study of 1476 patients showing congenital hand malformations. Macrodactyly is not inherited. Inglis (1950) listed 3 possible causes. (a) Abnormal blood supply, (b) Abnormal nerve supply, (c) Abnormal humoral mechanisms. Evidence suggests that nerves exhibit some control over tissue growth (Moore, 1972). So Inglis suggested that impaired nerve function stimulates uncontrolled growth of other tissues. Streeter (1930) attempted to explain the defect on the basis of vitality of germplasm in the developing limb. Barsky (1967) explained the findings as resulting from a disturbance in the growth limiting factor that allowed the overgrowth to proceed. Wartheman uses the term macrodystrophia lipomatosa to describe the fibrous and lipomatous infiltrations of the subcutaneous tissues and of the median nerve. Johnson and Bonfiglio (1969) used the term lipofibromatous hamartomas to describe an overgrowth of the peripheral nerve accompanied by limited cutaneous or skeletal overgrowth. Paletta and Rybka (1972) reported on hamartomas of the median nerve associated with digital overgrowth, and Yeoman (1964) described several similar cases. Edgerton andTuark (1974) outlined the similarity between neurofibromatosis and macrodactyly in some cases though other cases show gigantisms without signs of neurofibromatosis. Kelikian (1974) believed that the most common variety of macrodactlyly should be called nerve territory oriented macrodactyly (NTOM). This allowed the association of overgrowth and nerve anomalies without specifying the cause and the effect. It would include both idiopathic gigantism and neurofibromatosis. Frykman and Wood (1978) used histologic grounds and inheritence to separate those cases of overgrowth associated with large quantities of fat intimately mixed with nerve fibres from cases of neurofibromatosis.

McCaroll reported on several cases of macrodactyly in which he completely excised the digital nerve. Tsuge (1967) believed the enlarged nerves to be the causative factor and hence excised the nerve branches. Paletta (1972) underwent the

same procedure and found no subequent overgrowth in long term follow-up.

Case Reports

The study was carried out in Medical College Hospital, and included 6 cases between 4 years to 14 years of age.

Case — 1 : A 4 year old girl was admitted with a huge overgrowth of the left 2nd and 3rd toes. There was also syndactyly between those two affected toes. The child was otherwise healthy but her blood pressure was 138/60 mm of Hg., which was rather high for her age.

Other investigations were normal. X-ray of the affected foot showed increase in the soft tissue shadow of both the affected digits, the metatarsal bones and all the phalanges of the affected digits were enlarged. Angiogram (Femoral) revealed hyperdynamic circulation ; the dye was traced till the metatarsal arteries and then could not be visualised further. This showed that (i) There was no abnormality with the basic arteries of the foot, (ii) The overgrowth of the digits was not due to any vascular mass, (iii) The rapid runoff of the dye beyond the metatarsal arteries leading to nonvisualisation of the vascular pattern distally and the hyperdynamic and circulation seen during angiography could be due to the presence of A-V fistulae which cannot be proved directly (Fig. 1-3),

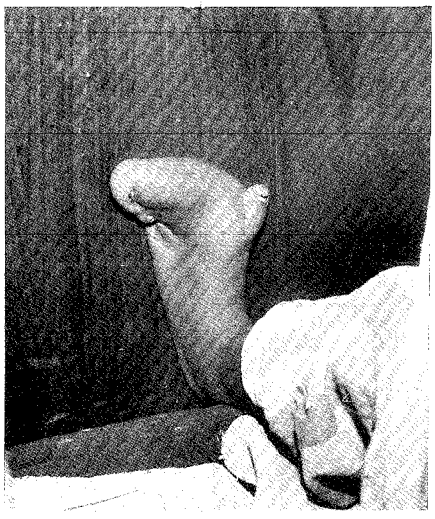

Fig. 1. Macrodactyly and Syndactyly affecting the second and third toes of the left foot.

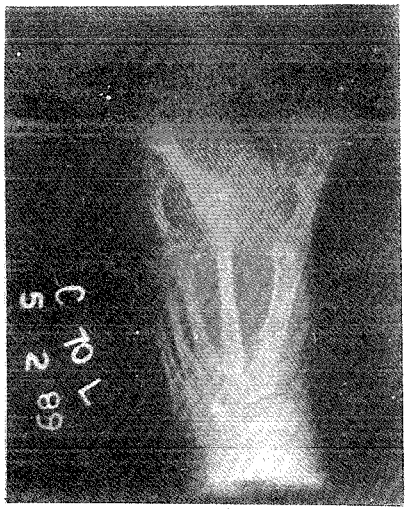

Fig. 2. Xray of the affected foot showing increase in size of the bones of the affected toes as well as the corresponding metatarsal bones.

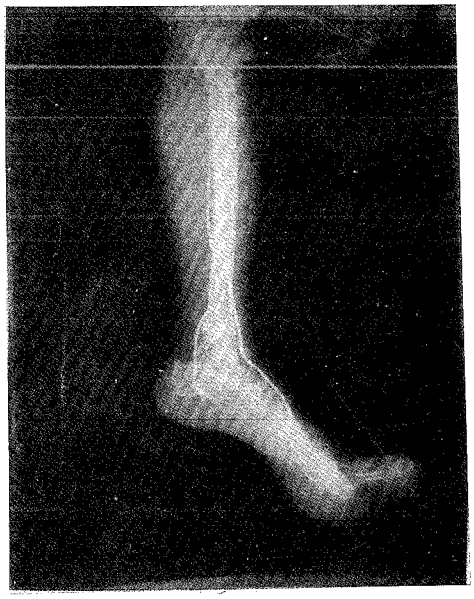

Fig. 3. Angiogram of the affected foot showing normal arterial pattern visible till the metatarsal arteries.

except by digital substraction angiography for which facilities are available only at very few centres.

The patient was put up for operation. Through zig-zag incisions the soft tissue underlying it was shaved, distal phalanges were excised and skin closed directly after excising the excess. However, some portions of the overlying skin got necrosed,

and secondary split skin grafting had to be done later. The Pathologist reported the excised mass as containing hypertrophied vascular, nervous, fibrous and fatty tissue. There has not been any significant growth of the digits since the operation which was done about six months back.

Case-2 : A 6 year old girl was admitted with recurrent overgrowth of the right 4th and 5th toes. She had been operated upon one and a half years back when distal phalanges of both the toes were excised and soft tissue shaving was done. This girl had a blood pressure of 140/60 mm of Hg. Other investigations were normal. X-ray showed overgrowth of the 4th and 5th metatarsal bones and angiographic studies were exactly like those of the previous case. Soft tissue shaving was done and the report showed it to contain only fibro fatty tissue. She had no post-operative complications. (Figure. 4-6).

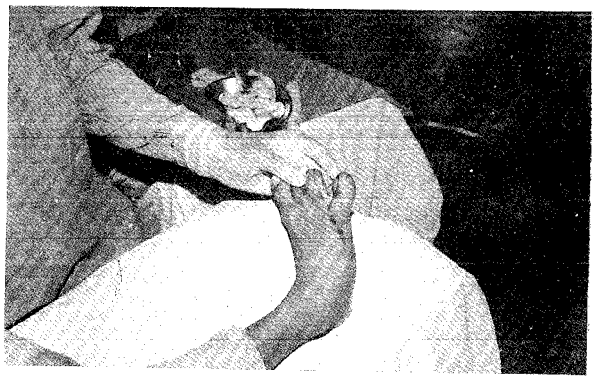

Fig. 4. Marking for the Zig-Zag incision.

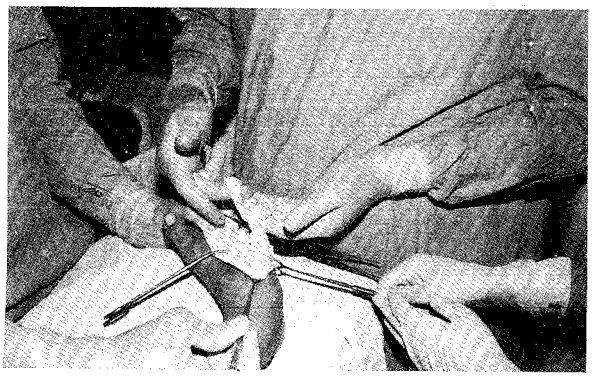

Fig. 5. Excess soft tissue displayed.

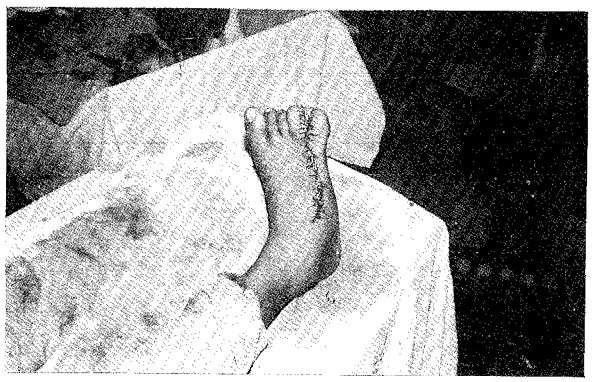

Fig. 6. After completion of the operation.

Case-3 : A 6 year old boy presented with huge overgrowth of the left index and middle fingers. His blood pressure was recorded to be 148/62 mm of Hg. X-ray of the part showed massive overgrowth of the phalanges of the affected digits, the corresponding metacarpals being thickened also. Angiographic studies revealed hyperdynamic circulation, and a rapid run off of the dye beyond the metacarpals. It was decided that soft tissue and bony excision will still leave a more than normal digit, thus amputation of the middle finger from the metacarpo-phalangeal joint was carried out.

Case-4 : A 14 year old girl presented with syndactyly and overgrowth of the 2nd and 3rd toes of the left foot. Soft tissue excision and excision of the distal phalanges was done 3 years earlier but the growth continued. Result of angiographic studies were like the previous cases and her blood pressure was 136/68 mm of Hg. Excision of soft tissue and the heads of the metatarsal bones were done.

Case-5 : A 12 year old boy presented with a moderately big 2nd toe of the right foot. Soft tissue excision was done on it one year back, but the growth continued. The BP was 130/60 mm of Hg. Angiographic findings were as in the previous cases. Soft tissue excision and excision of the middle phalanges of the affected toe was done. Splinting was done with a K-wire. Portion of the overlying skin sloughed out on the 7th post-operative day. Split skin grafting was done and K-wire was removed after 3 weeks.

Case-6 : A 2½ year old girl was admitted with overgrowth on the left second toe. Her blood pressure was 136/62 mm of Hg, and angiographic findings similar to the previous cases. Soft tissue

and middle phalanx of the toe was excised and splinted for 3 weeks. She had an uneventful post-operative period.

Discussion

Macrodactlyly is a rare condition. The incidence has been described as 0.9%. It is not inherited (Barsky, 1967 and Kelikian, 1974).

In the present series, females are affected a little more frequently than males (4 : 2); Syndactyly is present in 2 of the 6 cases (33%) — as opposed to published incidence of 10% (Flatt, 1977). In St X-ray of the part, the metacarpal and metatarsal bones of the corresponding digits were definitely involved and enlarged as opposed to Mustarde's report, that they do not. Almost all the patients had somewhat high systolic pressure (for their age). Angiographic study revealed the possible existence of single/multiple arteriorvenous communications. This could be the initiating factor for the gigantism.

We have observed that all structures of the involved part including skin, subcutaneous fat, nerves, arteries and bone are affected in the overgrowth. The skin is thickened, increased subcutaneous fat appears with varying amounts of fibrous tissue stroma, digital arteries are enlarged and have large lumina, normal appearing flexor tendons pass through thickened flexor tendon sheath, digital nerves are thickened, tortuous and greatly enlarged, the bony trabeculae are well formed with an increased amount of normal appearing fatty marrow. The severity of involvement varies from a symmetrically enlarged digit with minimal increase in length and circumference to grossly gigantic fingers with angulation, ankylosis and decreased sensibility causing grotesque deformities (Millesi, 1974).

Treatment of the condition remains unsatisfactory. Soft tissue excision en bloc was done in all cases, being repeat procedures for some. Epiphyseal arrest procedures were not done in any case, because though often successful in limiting longitudinal growth, the bone usually enlarges circumferentially from onlay bone formation. A number of techniques are available to reduce the bone size, all of them creating a smaller digit with sacrifice of the joint movement occasionally. We have excised the proximal phalanx in some, the distal phalanx in others and even the head and neck of the proximal phalanx in one. With markedly distorted and grotesque deformities, amputation is the procedure of choice. Most patients show poor wound healing, and in our series, we had to resort to secondary skin grafting because of loss of skin, in two patients. Remarkably, the resultant sensory deficit in all the cases has been less than expected.

Conclusion

Six cases of macrodatyly have been investigated and operated, and the excised tissue hislogically studied. The relevant literature has been reviewed.

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