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Macrodactyly of the Great Toe: A Rare Case Report

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Abstract

Macrodactyly is a rare congenital condition which can involve hands or feet. It is characterized by overgrowth of bone and soft tissues. Macrodactyly is not cancerous. Other names for this condition are local gigantism, megalodactyly, localized hypertrophy.

Keywords: Etiology, Hypertrophy, Lower Limb, Peripheral Nerve, Tendons

Introduction

The term macrodactyly means large digit. Macrodactyly is an uncommon congenital condition which can involve upper or lower limb and presents as an overgrowth of entire digit. This can affect one or multiple digits, unilateral or bilateral. Upper limbs are more commonly affected than lower limbs. According to Barsky's definition[1] of macrodactyly phalanges, tendons, nerves, subcutaneous fat, nails, skin all must be enlarged[2]. Etiology of this condition is largely unclear, but according to some studies it is caused by abnormal neural control of sensory distribution in a peripheral nerve[3].

Macrodactyly of foot is usually an isolated entity. Patient with foot macrodactyly commonly presents with complaints[4] of difficulty in walking, gait disorders, cosmetic concerns, inability to fit in normal shoes, stiff toes, nail changes, distal ulcers, osteomyelitis, aesthetic problems. This condition is emotionally & physically debilitating; generally multiple surgical interventions may be needed. The goal of surgical treatment is to achieve an aesthetically & functionally acceptable limb[5].

Case Presentation

A 35 years old female patient from Kota, Rajasthan presented with enlargement of great toe of left foot. Her toe was enlarged since birth and had been continuously progressing in size. Clinical examination showed enlarged great and stiff toe with deformity. On further history taking she denied any pain, tenderness, numbness in toe but reported that the condition affected her daily activities.

Figure 1: Enlarged Left Great Toe, Note The Nail Deformity And Lateral Deviation Of Other Unaffected Digit [Frontal And Lateral View]

Radiographs of foot showed enlarged soft tissue and bony components of left great toe.

Deformity of first metatarsal and phalanges were revealed. Rest other bony structures were grossly normal. Diagnosis of macrodactyly was confirmed by clinical and radiological findings.

Figure 2: X Ray of Left Foot Showing Enlarged And Deformed Great Toe With Soft Tissues And Bony Overgrowth [Note Abnormal Fusion of First Interphalangeal & MCP Joint With Banana Deformity Of Great Toe]

Patient was planned for surgery after proper pre anesthetic checkup. Under spinal anesthesia great toe amputation was done. Cut section of amputated part showed soft tissue and bony enlargement. Specimen was sent for histopathological examination which showed excessive proliferation of subcutaneous adipose tissue, increased fibrous septa with thinning of epidermis with enlarged bony components.

Post-operative period was uneventful. Patient was given iv antibiotics and pain killers. Wound cleaning and dressings were done regularly.

Figure 3: Amputation of Great Toe [Note Presence of Excessive Soft Tissue and Bony Components]

Figure 4- Post Amputation [CRD (Corrugated Rubber Drain) Placed Along Suture Line to Evacuate Any Collection]

Discussion

Pathogenesis of macrodactyly-

- Lipomatous degeneration (macrodystrophia lipomatosa progressiva)[7,9]

- association with neurofibromatosis
- nerve dysfunction
- abnormal growth factors(in utero disruption of growth limiting factors).

Hardwicke et al.[8] described four types of macrodactyly – type I, macrodactyly with lipofibromatosis of a nerve, either of a static or progressive subtype; type II, associated with neurofibromatosis; type III associated with hyperostosis; and type IV, associated with hemihypertrophy.

Classification of macrodactyly[10]

1. True and False macrodactyly-True macrodactyly involves hypertrophy of all the structures of the digit i.e. skin, toenail, subcutaneous fat, bone, nerves and blood vessels. False macrodactyly presents as hypertrophy of predominantly one tissue type.
2. Static and progressive type-Static macrodactyly involves digits that are enlarged at birth and grow proportionately with rest of hand and foot. In progressive macrodactyly digits grow at a faster rate than the other unaffected digits becoming significantly larger over time.
3. Primary and secondary macrodactyly-In primary condition overgrowth of affected digit occur without any associated limb enlargement. On other hand secondary macrodactyly is associated with some underlying syndromes eg. neurofibromatosis, proteus syndrome, Maffucci 's syndrome, Ollier's disease, Klippel Trenaunay syndrome, vascular malformation.

Management of macrodactyly

Conservative methods are of limited value. Surgical procedures include,

- Tissue debulking procedures -most commonly attempted procedure and usually done with other interventions.
- Phalangectomy- involves shortening of affected toe. Barsky combined it with arthrodesis.
- Digit shortening
- Amputation -usually done in metatarsals involvement.
- Osteotomy
- Surgery of first Ray- not done routinely due to importance of great toe during gait and cosmesis.
- Epiphysiodesis – used to arrest growth mainly in hallux in young age.

Conclusion

This case report focuses on presenting a rare condition that is usually underdiagnosed. Early diagnosis and correct treatment at appropriate age can avoid radical measures like amputations.

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