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MACRODACTYLY- A Case Report

Case Report

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Abstract:

A case of macroductyly of the right index finger in a male aged eleven years is reported here. This subject was found to have abnormally large index finger in his right hand. This case was not associated with any other physical abnormality. Other fingers and toes were normal. This boy was a second child born to a consanguineous couple. His siblings did not have any anomalies. The macroductyly of the index finger in this case did not cause difficulty in carrying out day to day activities like writing, lifting heavy objects and also for using the hand to have food. Such cases may have some morphological and associated genetic importance.

Macroductyly is a rare congenital anomaly in which there is enlargement of one or several digits of hands or feet hence this report.

Key words: macrodactyly, dactylomegaly, macromelia, hemihypertrophy, megalodactyly, digital gigantism, syndactyly, macrosomia, digital anomalies, lipofibromatous hamartoma

Introduction:

Macrodactyly(dactylomegaly, digital gigantism) is a rare congenital anomaly in which there is enlargement of one or several digits of hands or feet(1). The overgrowth is limited to or predominantly affects the digits and should be distinguished from more extended malformations such as macromelia or hemihypertrophy. Sometimes the words megalodactyly or digital gigantism is used synonymously. Macrodactyly does not appear to be an inherited condition. Although its cause is uncertain, three possible factors are strongly suspected: abnormal nerve supply, abnormal blood supply and abnormal humoral mechanism(2). Macrodactyly most commonly exists without other conditions but syndactyly is associated with macrodactyly in about 10% of patients(3). Digital gigantism is an unusual congenital anomaly that is present at birth or recognized in early infancy and is progressive during the period of normal skeletal maturation. It involves cell types that are predominately affected by neurogenic growth control and follow a distribution similar to the sensory supply of a major peripheral nerve, usually the median nerve(4). There seem to be three variants that overlap somewhat in clinical presentation. They are type I digital gigantism with lipofibromatous hamartoma of a peripheral nerve, type II digital gigantism associated with neurofibromatosis, and type III hyperostotic digital gigantism.

Macrodactyly in the majority of patients is an isolated finding without other associated symptoms or systemic involvement (1). There is a range of other, mostly congenital pathologic conditions in which localized overgrowth may mimic the clinical picture of macrodactyly.

These include neurofibromatosis, primary lymphatic disorder (Milroy disease) and vascular malformation, for example Klippel-Trenaunay-Weber syndrome(5). Several rare hereditary syndromes include hamartomatous changes that possibly present as macrodactyly, for example, Proteus syndrome, Bannayan Syndrome, Maffucci Syndrome and Ollier disease. Diagnostic aids in cases of macrodactyly should include radiography and computed tomography to reveal the extent of bone and tendon involvement. MRI is recommended when the clinical features and plain film findings are indeterminate. Angiography and lymphangiography should be performed when vascular or lymphatic changes, respectively are clinically suspected. Biopsies can provide information on the tissue elements that take part in the formation of the hamartoma.

The patient in this case report was an 11 yr old boy who was born with abnormal enlargement of the right index finger. The enlargement has not adversely affected his normal functions as he has been able to write and perform other functions with the same hand. He was seen at the St Joseph's hospital Kingston Jamaica on the 9th of July 2016 accompanied by his mother having come for a school medical and I observed the gigantic size of the right index finger. The right index finger was 20% longer(6cm vs7.2cm) with the diameter up to 300% larger than the corresponding finger on the left at the middle of the distal phalanx(1.8cm vs 5.4cm)Fig 1, & 2 confirmed by Xray Fig 3)

Surgical treatment

Treatment should be individualized and initiated early in life. Epiphysiodeses, osteotomies, and division of branches of the supplying digital nerves may be indicated during childhood. In the adult, arthrodesis of the interphalangeal joints with bony shortening or ray resection may be indicated (6). Skin flap necrosis is a common complication postoperatively.

No treatment is being contemplated in this case as the deformity does not affect the normal function of the hand and the patient/parents are not interested in cosmetic correction.

Discussion:

Macroductyly(Digital gigantism) is an unusual congenital anomaly that is present at birth or recognized in early infancy and is progressive during the period of normal skeletal maturation. It involves cell types that are predominately affected by neurogenic growth control and follow a distribution similar to the sensory supply of a major peripheral nerve, usually the median nerve. There seem to be three variants that overlap somewhat in clinical presentation. They are type I digital gigantism with lipofibromatous hamartoma of a peripheral nerve, type II digital gigantism associated with neurofibromatosis, and type III hyperostotic digital gigantism. Treatment should be individualized and initiated early in life. Epiphysiodeses, osteotomies, and division of branches of the supplying digital nerves may be indicated during childhood(7). In the adult, arthrodesis of the interphalangeal joints with bony shortening or ray resection may be indicated. Skin flap necrosis is a common complication postoperatively

Images



Image 1

Image of patient's hand showing enlargement of right index finger



Image 2
The same hand with the dorsal view of the digital gigantism

Figure 3
Xray of the hand showing enlargement of the phalanges involved



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