

Answer to Dermacase

continued from page 171

2. Megadactyly

Megadactyly is a rare congenital anomaly characterized by disproportionate hypertrophy of 1 or more digits of the hands or feet. The incidence of megadactyly is estimated to be 1 in 3000, with equal likelihood of hand or foot involvement and a slight male predominance.^{1,2} True megadactyly entails enlargement of all components of the involved digits (bone, nerve, vessels, fat, skin, etc).¹ The anomaly has also been called *digital gigantism*, *dactylomegaly*, *megalodactyly*, *macromelia*, *megalomelia*, and (incorrectly) *macrodistrophia lipomatosa*.³ The cause and pathophysiology of megadactyly are unclear; however, it has been postulated that the condition is due to abnormalities in nerve innervation or blood supply to the affected area.⁴ Isolated megadactyly is not known to be an inherited condition.¹

Clinically, megadactyly can present as an isolated phenomenon (as in the present case) or as part of a generalized condition such as neurofibromatosis, hemangiomas, or Klippel-Trenaunay-Weber syndrome, among others. For both hands and feet, the second and third digits are frequently involved.^{2,5} Two general types of megadactyly have been described. Static megadactyly is present at birth, with hypertrophy of the affected digits taking place in proportion to the normal growth of the unaffected digits. Progressive megadactyly typically presents in early childhood and is characterized by rapid and disproportionate hypertrophy of the affected digits, often with the development of angular deformity.^{1,4}

Differential diagnosis

Because of its possible association with various genetic syndromes, the differential diagnosis of megadactyly should include neurofibromatosis, Klippel-Trenaunay-Weber syndrome, Beckwith-Wiedemann syndrome, Milroy disease, and Proteus syndrome.⁶ In newborns or young infants, isolated megadactyly should be a diagnosis of exclusion. If the patient is presenting at birth, a detailed physical examination and family history are essential to determining the presence of other signs or symptoms of



systemic involvement. Referral to a medical geneticist is warranted if megadactyly presents in the context of a greater syndromal picture. The diagnosis is more straightforward in older children who present as otherwise healthy.

Treatment

In cases of isolated megadactyly, treatment is indicated to relieve pain, poor foot function, or poor shoe fit.^{4,5} Cosmetic improvement is important, but secondary. Treatment is aimed at producing a painless foot that can accommodate regular shoes. There are no satisfactory non-surgical treatments for megadactyly.⁴ Surgical interventions include soft tissue debulking (with or without osteotomy), epiphysiodesis, phalangeal amputation, and metatarsal or phalangeal ray resection.^{2,4,5} Evidence is mixed as to the best surgical approach; however, ray resection has been shown to produce the best outcomes in cases with metatarsal involvement.⁷

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Competing interests

None declared

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