Hereditary Pincer Nail

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Pincer nail is a rare dystrophy characterized by transverse overcurvature that increases distally along the longitudinal axis of the nail. Etiology, pathogenesis, and inheritance of this dystrophy are not clear. We report on 3 remotely consanguineous siblings (2 girls, 1 boy) with pincer nail of the toes. Case distribution in this family raises the possibility of autosomal-recessive mendelian inheritance.

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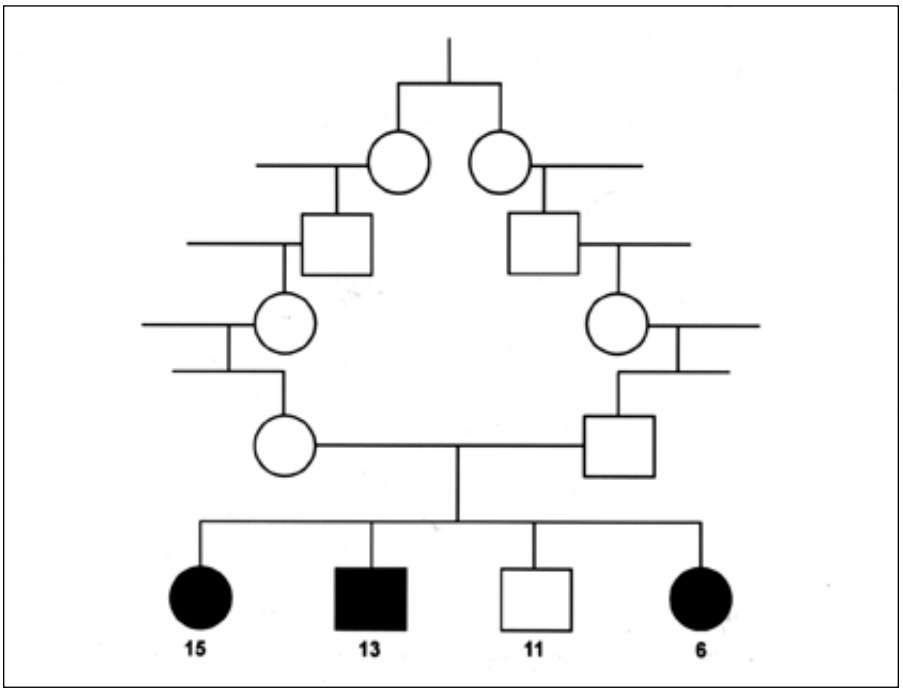

Figure 1. Autosomal-recessive inheritance can be inferred from the pedigree of this family with pincer nail. White squares indicate unaffected males; black square, affected males; white circles, unaffected females; black circles, affected females.

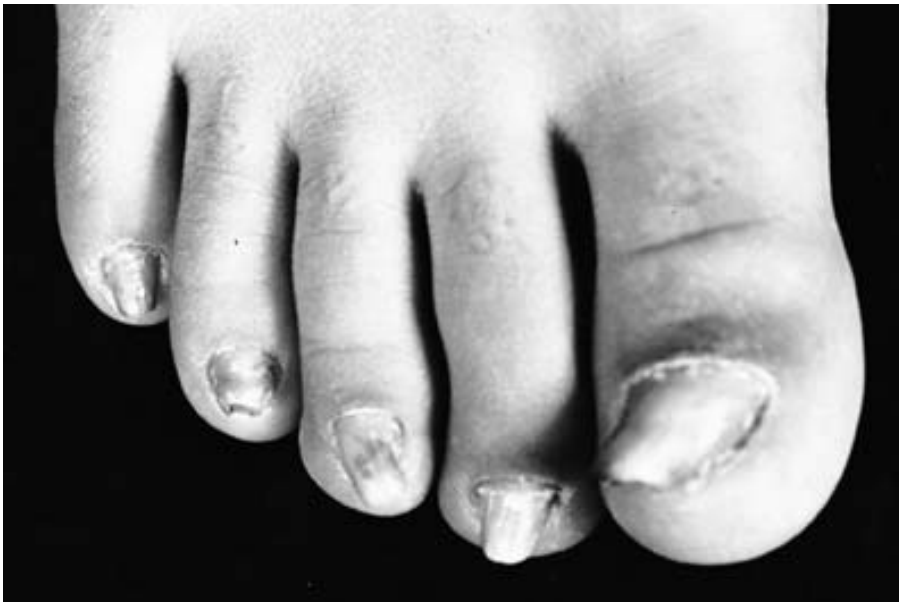

Figure 2. Foot of 6-year-old girl with pincer nail dystrophy of all digits.

Pincer nail is a rare dystrophy characterized by transverse overcurvature that increases distally along the longitudinal axis of the nail. The digit becomes pinched, and soft tissue is lost. Most cases are sporadic and considered to result from a developmental anomaly. Nevertheless, the condition also has been reported as being acquired from mechanical deformation of the nail because of subungual exostosis, local trauma, onychomycosis, epidermoid cyst, or psoriasis.[1-3] Recently, it has been reported after treatment with β-blockers and as a sequela of Kawasaki disease.[4,5] Chapman[6] suggested an autosomal-dominant inheritance of this phenomenon when he first described direct transmission through 3 generations. We report distribution in a family suggestive of autosomal-recessive mendelian inheritance.

Case Reports

Three siblings in a family with remote consanguinity (family tree in Figure 1) presented to our clinic with asymptomatic pincer nail of the toes. The

patients, a 6-year-old girl (Figure 2), a 15-year-old girl, and a 13-year-old boy, had had pincer nails of all the toes since early infancy; a second brother, the parents, and other family members had normal nails. There were no histories of trauma. Results of complete physical examinations were unremarkable except for excessive transverse nail curvature at the distal end of the toes. Onychomycosis was excluded clinically and by culture. Radiologic examination of the fingers showed no distally directed exostoses.

Comment

Etiology and pathogenesis of pincer nail are not fully understood. In 1973, Chapman[6] described several cases of pincer nail in 3 consecutive generations of a family. In each case, disease onset occurred in early adulthood (ages teens–early 20s). This pattern strongly suggests that the condition might be inherited as an autosomal-dominant mendelian characteristic. In our report, 3 remotely consanguineous siblings had pincer nail dystrophy. That 3 of 4 children had this dystrophy and that the rest of the family had normal nails are highly suggestive of autosomal-recessive inheritance. In addition, in each of our cases, disease onset occurred in early childhood. The different mode of inheritance suggests that the disease may have an autosomal-dominant variant, expressed in early adulthood, and an autosomal-recessive variant, expressed in early childhood.

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