Bilateral Triphalangeal Thumbs

A 10-year-old boy was brought to the outpatient department with the complaint of alopecia areata. The appearance of his hands was striking. Both hands appeared to have no thumb and five completely normal looking fingers arising from a triangular shaped palm (*Figs. 1,2*). Left great toe was also broad and flattened. A complete physical examination revealed no other bony abnormality, no pallor and a normal systemic examination. Child was developmentally normal and was able to write well. There was no family history of any bony anomaly.

Triphalangeal thumb (TPT) is a rare congenital disorder characterized by a long, finger-like thumb with three phalanges instead of two. It can occur as an isolated defect, in association with other abnormalities of the hands and feet, or as a part of a syndrome like TPT-polydactyly syndrome, tibial hemimelia-polysyndactyly-TPT syndrome, acropectoral syndrome, Aase syndrome (TPT with congenital hypoplastic anemia), Townes-Brocks syndrome (TPT, deafness, anal anomalies), Townes-Brocks syndrome with renal dysplasia, Laurin-Sandrow syndrome (underdeveloped nasal bones, large heads of mandibular condyles, synostosis of tarsal bones along with polysyndactyly and TPT), a syndrome of TPT, Glanznann's thrombasthenia and deafness and in few patients with Poland-Moebius syndrome. TPT has also been reported in association with psychomotor retardation. Families have been reported in literature in which one member may have isolated TPT while other members may have TPT along with other malformations mentioned above. This is due to the pleiotropic expression of the genes.

Isolated TPT anomaly has been mapped to chromosome region 7q36. Sporadic cases have been described, but it is usually inherited as an autosomal dominant trait. This condition has its embryological basis in the abnormal development of the apical ridge ectoderm during the fifth week of embryogenesis, which plays a role both in early growth of limb and definition of its skeletal elements. Inadequate induction of growth and

Fig. 1. Palmar aspect of both hands showing triphalangeal thumbs with triangular shape of palms.

Fig. 2. Dorsal aspect of both hands showing triphalangeal thumbs.

differentiation on the preaxial side of hand can produce a spectrum of conditions ranging from complete thumb aplasia to varying degrees of hypoplasia to a relatively long triphalangeal thumb lying in the same plane as the fingers. The muscle mass making the thenar eminence is also diminished and the palm appears triangular.

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