PALMAR DERMATOGLYPHICS IN MARFAN'S SYNDROME

S Premalatha, K Kannaki and P Sukumar

Palmar dermatoglyphics were studied in two typical cases of Marfan's syndrome by using the standard ink and roller method. Simian crease, extra-digital transverse crease, increased atd angle, high position of the axial triradius and white lines were recorded.

Key words : Dermatoglyphics, Marfan's syndrome.

Marfan's syndrome belongs to the group of dys-elastoses and exhibits musculo-skeletal, ocular and cardio-vascular manifestations. This condition is transmitted by an autosomal dominant gene but sporadic mutants are recognised.[1]

A dermatoglyph is a natural carving in the skin that is a furrow or a fold.[2] The development of dermatoglyphic pattern in utero is strongly influenced by genetic factors,[3,4] and it has been studied in various genetically determined disorders.

A study of palmar dermatoglyphics was done in two female cases of Marfan's syndrome and on 10 phenotypically normal females, taken as controls.

Case Reports

Case 1

A 13-year-old girl, born to non-consanguineous parents, attended for tall stature, long thin limbs, lax skin and hyper-extensibility of the joints. Her arm span was 140 cm which was slightly less than her height (142.5 cm). Hyper-extensibility of the joints also was present. Steinberg's sign was present—when the thumb was enclosed in the closed fist, it protruded beyond the medial border of the hand. Cardio-vascular, respiratory, gastro-intestinal and genito-urinary systems were clinically normal. There was no ocular involvement. She developed psychiatric manifestations for which treatment was given. Two siblings, one elder brother and one elder sister also were having tall stature with long thin limbs. Parents and 4 other siblings were normal. The heights of father and mother were 162.5 cm and 155 cm respectively.

Case 2

A 12-year-old girl, born to consanguineous parents presented with tall stature, long thin limbs, hyper-extensibility of the joints and a small firm nodular lesion on the second toe on the right side. She had congenital genu recurvatum with hyper-extensibility of the left knee joint which was corrected. She had long facies, lax skin, blue sclera, microcornea and high-arched palate. Her height and arm span were equal (142 cm). Steinberg's sign was positive. Cardio-vascular, respiratory, gastro-intestinal and genito-urinary systems were clinically normal. She was anaemic. X-rays of the hands and feet showed elongated metacarpals and metatarsals respectively in addition to the elongated phalanges. The metacarpal index was 9.63 (normal 5.4-7.5). Calcium deposit was observed in the right foot in the skiagram. Excision biopsy of the nodular lesion of the right fourth toe revealed calcinosis cutis. Serum calcium and serum phosphorous levels were normal, but serum alkaline phosphatase level was slightly increased. Parents were of normal height. The heights of father and mother were 160 cm and 150 cm respectively. One male sibling, elder to the proband was normal.

From the Department of Dermatology, Madras Medical College and Government General Hospital, Madras-600 003, India.

Address correspondence to ; Dr. S. Premalatha, D. 111. C., MIG Flats, Sangeetha Colony, Ashok Pillar Road, K. K. Nagar, Madras-600 078.

Dermatoglyphic pattern

On the finger-tips, the ulnar loops were seen in 65% and whorls in 35%. Arches and radial loops were not observed. The patterns were absent in the thenar (Th) and interdigital (I_1, I_2 and I_4) areas of the palms; I_3 area however, showed patterns in the left palm in both the cases. Case 1 showed a small loops (l) and case 2 showed a large loop (L). Case 1 showed a radial loop pattern (L^r) in the hypothenar area of the right palm. White lines were seen in both cases. Extra-digital transverse creases were observed over the middle phalanges of all the fingers and distal phalangeal regions of the middle and index fingers on the right hand in case 1 (Fig. 1). Simian crease was well observed in the right palm in case 2 (Figs. 2 and 3).

The mean TFRC was 118 compared to 144.5 in the controls. The mean atd angle was 61° which was more when compared to that of

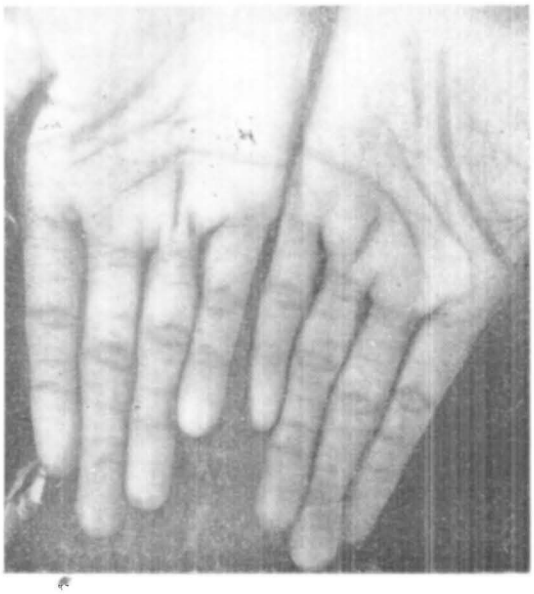

Fig. 1. Extra-digital transverse creases over the middle phalanges of all fingers and the distal phalanges of the index and middle fingers of the right hand (Case 1).

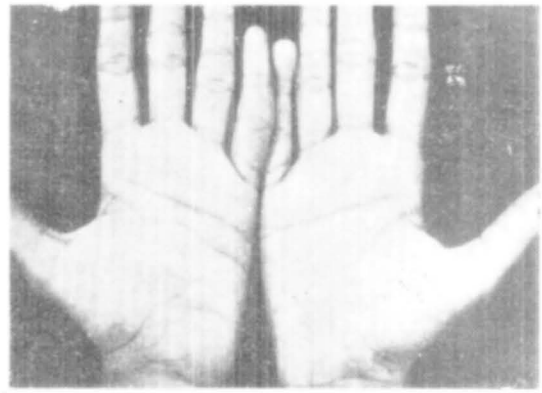

Fig. 2. The elongated palms with Simian crease in both palms (Case 2).

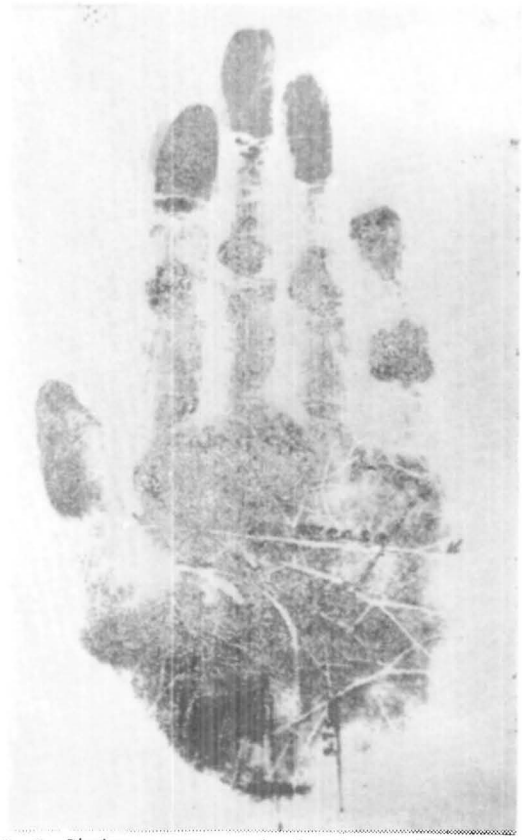

Fig. 3. Simian crease, maximal atd angle (87°) and position of axial triradius almost at the centre of the palm-t" position.

control subjects (42.5°). The mean a-b ridge count was similar to that of controls (38.05 in control subjects and 38.5 in Marfan's syndrome). The position of axial triradius was t in case 1 and t" in case 2 (Fig. 3).

Dermatoglyphic study of the available family members of case 1 revealed the presence of extra-digital transverse creases over the middle phalanges of index, middle and ring fingers of both hands of her elder brother and a whorl pattern in the hypothenar area of the right palm of her elder sister.

The father of case 2 had well marked extra-digital transverse creases over the middle phalanges of index, middle and ring fingers of the left hand.

Comments

Two typical cases of Marfan's syndrome were studied along with dermatoglyphic findings. As calcinosis cutis has been usually reported in elastic tissue disorders such as pseudoxanthoma elasticum and Ehlers-Danlos syndrome, it is a point of interest to record this occurrence in a case of Marfan's syndrome (Case 2).

We have observed interesting dermatoglyphic features such as simian crease, extra-digital transverse crease, increased atd angle, high position of axial triradius and white lines in two cases of Marfan's syndrome. In addition, extra-digital transverse crease has been observed over the middle phalangeal regions in some of their family members who were phenotypically normal. The authors would like to stress that more cases of Marfan's syndrome have to be studied in future for such dermatoglyphic findings to find out their significance and importance in the field of dermatoglyphics.

References

1. Barr DGD and Forfar JO : Disorders of bone and cartilage, in : Text Book of Pediatrics, 2nd Ed, Editors, Forfar JO and Arneil GC : Churchill Livingstone, London, 1978; p 1344-1402.
2. Gibbs RC : Fundamentals of dermatoglyphics, Arch Dermatol, 1967; 96 : 721-725.
3. Schaumann B and Milton A : in Dermatoglyphics and Chromosomal Aberrations, Human Chromosome Methodology, 2nd ed, Editor, Yunis JJ, Academic Press, New York, 1974; p 271-310.
4. Polani PE and Polani N : Chromosome anomalies, Mossaicism and dermatoglyphic asymmetry, Ann Human Genet, 1969; 32 : 391-402.