

Case Report

The enigma of the simian crease: case series with the literature review

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ABSTRACT

Simian crease (SC), a single transverse crease that extends from across the palm of the hand. The presence of a single transverse palmar crease or the SC can be, but not always, a symptom associated with abnormal medical conditions. In dysmorphology, the clinical significance of variability of palmar creases especially SC are widely recognized and have occurred more frequently in patients with various congenital anomalies that are caused by genetic as well as environmental insults in the first half of pregnancy. SC has been studied extensively in children especially with Down's syndrome (DS) where the incidence of SC is more than 63%. In this case report, two families are discussed with relevance to the presence of SC. The first family with two children with DS have no SC in any of them, whereas the second healthy family only the father who is a doctor by profession having an SC.

Keywords: Dysmorphology, Palmar creases, Simian crease, Downs syndrome

INTRODUCTION

Palm creases have wide practical use because they can be analyzed and interpreted quickly without any physical pain, high costs or age considerations.¹ Simian crease (SC) are the single horizontal crease that extends across the palm of the hand in human beings. It is also called a four-finger crease, single palmar flexion crease or a single upper palm crease. Because it resembles the usual condition of non-human simians, it is also known as a SC or simian line.² As palm creases and SC, in *per se*, are helpful in discovering anthropologic characteristics and diagnosing several diseases, including chromosomal aberrations, they have been analyzed qualitatively² and quantitatively.³ Larger studies were done in children with cytogenetically confirmed patients of Down's syndrome (DS) and SC is common in DS.⁴ As DS is commonly seen, in any child with a SC the first premonition is to look for any signs of trisomy 21.⁵ In this case report, two families are reported to highlight the significance of SC in the population.

CASE REPORTS

Case 1

The first family includes two children with DS, born of non-consanguineously married parents attending outpatient clinic in the Department of Paediatrics, Amala Institute of Medical sciences. Parents were healthy and did not have SC in their palms (Figure 1).

Father was 42 and mother 31 years when she delivered the first girl child. No antenatal issues for the parents. Perinatal period was uneventful. First child, now 12 years old, was detected to have hypothyroidism and were on thyroxin. She has mental retardation, detected to have heart disease in childhood during evaluation for recurrent respiratory tract infection. She was diagnosed to have atrial septal defect and device closure was done in 2013. On general physical examination, she has the mongoloid slant, clinodactily, sandal gap, high arched palate. SC is not present. Systemic examination is normal now. Attending special school for mentally retarded children.

Father was 47 and mother 36 years when she delivered the second girl child. No antenatal issues for the parents. Perinatal period was uneventful. The second child is now 7 years old. She was also detected to have hypothyroidism and is on thyroxine. She has mental retardation and heart disease in childhood was detected during the evaluation for recurrent respiratory tract infection. She was also diagnosed to have atrial septal defect, and pericardial patch closure was done in 2013. On general physical examination, she has the mongoloid slant, clinodactily, sandal gap, high arched palate. SC is not present in her palms as in her sister. Systemic examination is normal now. She is also attending special school for mentally retarded children.

Case 2

A doctor couple attended the outpatient for immunization of their two children. The family was healthy, and parents were non-consanguineously married. The doctor was found to have an SC in his both palms. The wife was healthy and had no antenatal issues. The elder sibling was 10 years and the second sibling was 5 years old girl and both of them were healthy with no SC in either mother or two children (Figure 2). Systemic examination was normal.

DISCUSSION

Dermatoglyphics has an important role in dysmorphology and should be used in conjunction with the physical examination rather than as an independent diagnostic test⁶ when the patients present with chromosome abnormalities, aneuploidies and newly recognized deficiency and duplication syndromes. SC and the palmar flexion creases appear on the palms of the hands and soles of the feet and usually develop by the 12th week of gestation in the fetus.⁷ Even though the timing of development of the flexion SC has now been established, their origin is less well understood as per the literature. Both primary genetic determination and development secondary to flexion function have been well postulated as the mechanisms underlying the SC development. There is a close correspondence between the appearance of the flexion creases and hand and digital movement (which, in turn, is associated with joint formation and muscular function) that can be ascribed to these earlier factors.⁸ Hence, any insult occurring early in the fetal development can hamper the normal pattern of SC. It has been found that most creases including the SC develop concurrently with the appearance of the fetal volar pads and the rest develop independently of them and also reports are available to correlate that SC seem to develop independently of the flexion movements of hand and foot,⁹ whereas SC in a monkey is due to a particular movements such as the thumb flexion in the same direction as the other fingers when the monkey grasps a stick. As the hand movement of humans is freer than that of animals, palm creases of humans are more prominent than those of animals.¹⁰

Larger studies have been done in the population with SC and with particular reference to DS.^{4,11} Many methods have been postulated to diagnose SC in DS children.^{11,12} Single palmar creases are less common than two palmar creases,

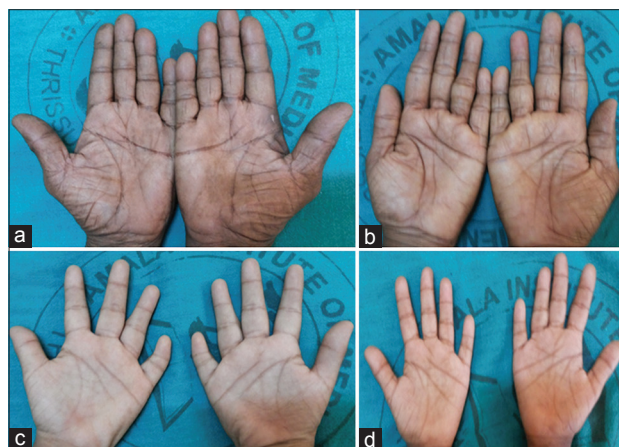


Figure 1: Photograph of palm of first family members (a) father, (b) mother, (c) elder sibling with Down's syndrome and, (d) younger sibling with Down's syndrome.

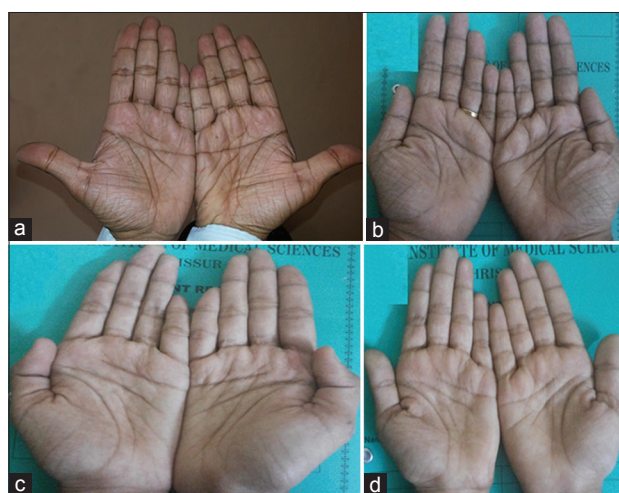


Figure 2: Photograph of palm of second family members, (a) father, a doctor with simian crease, (b) mother, (c) elder sibling and (d) younger sibling.

however 10% of the population have one SC on one hand and 5% have one SC on both hands.^{2,3,10} Males are twice more common than females to have this characteristic, and it tends to run in families.¹³ It is frequently seen among Asians and Native Americans than among other populations, and in some families there is a tendency to inherit the condition unilaterally.^{2-4,10,13} In our study, the doctor has SC on the both palms and is normal according to the various studies.¹³

Palm creases have additional features that might indicate the possibility of chromosomal aberrations.^{12,14-18} SC is frequent, and incidence is high in DS (trisomy 21).^{4,5,12,19} However in our children with DS, an SC was not present and so we have to look for other features of trisomy 21. Hence, SC should not be used as an independent test to diagnose DS.⁶ SC are also useful for diagnosing intrauterine toxin exposure,^{3,17} fetal alcoholic syndrome, mental retardation, neoplasia, and other diseases.^{14,19} A unilateral SC was also reported in the case of chromosome 9 mutation causing nevoid basal cell carcinoma syndrome and Robinow syndrome. It is also sometimes

found on the hand of the affected side of patients with Poland syndrome and craniosynostosis.^{14,15} Forbes reported SC in Klinefelter syndrome,¹⁶ pseudohypoparathyroidism¹⁶ and gonadal dysgenesis.¹⁶ Other syndromes with SC are Cohen syndrome, Turner syndrome, cri du chat syndrome (chromosome 5), Noonan syndrome (chromosome 12), Patau syndrome (chromosome 13), Edward's syndrome (chromosome 18), and Aarskog-Scott syndrome (X-linked recessive)^{11,15,16} and congenital rubella syndrome.¹⁸ The knowledge of dermatoglyphics is important and is an interesting part of science.²⁰ An infant with a single palmar crease may have other symptoms and signs that, when taken together, define a specific syndrome or condition.

Diagnosis of that condition is based on family history, medical history, and complete physical examination. So as a paediatrician it is pertinent to look for these and never miss them! This two case studies concluded that the first family with children with DS have no SC in any of them, whereas the second healthy family with only the father having a SC who is a doctor by profession with no SC in his two children and wife. These variations in the occurrence of SC in our population should warrant further studies.

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