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A and B preaxial polydactyly with syndactyly of feet and hands in the same person – a case report

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ABSTRACT

Preaxial polydactyly of the hand is more common than postaxial polydactyly and postaxial polydactyly of the foot is more common than preaxial in white patients. Syndactyly is commonly regarded as next in frequency among the congenital hand deformities. Preaxial polydactyly of the feet and hands and polydactyly type A and B with syndactyly in the same individual is a rare condition. In this work we present this rare disorder in male infant. The physical and X-ray examinations showed preaxial polydactyly type A of both hands and A and B polydactyly with syndactyly of feet.

Key words: congenital deformities, polydactyly, syndactyly

INTRODUCTION

Hand anomalies are often diagnosed during prenatal ultrasonography. The spectrum of malformations varies from subtle finger deformities to the complete amputation of limbs [1]. Polydactyly is the most frequent of congenital hand and foot deformities. Polydactyly occurs both in a sporadic form and in a hereditary form [2-6]. Each polydactyly was defined as a preaxial or postaxial and type A or B. Preaxial polydactyly of the hand is more common than postaxial polydactyly and postaxial polydactyly of the foot is more common than preaxial in white patients [3,7-10]. In polydactyly type A, the extra digits are well developed and they are joined to the normal fingers or an extra metacarpal bone. In polydactyly type B, there is no skeletal structure and the extra digit consists of a pedunculated skin tag. Classification of syndactyly contains type I- fingers are connected by skin and type B- webbing of fingers including bone structure. Syndactyly can be complete or partial, when usually only the bases of the fingers are joined. Preaxial polydactyly of the feet and hands and polydactyly type A and B with syndactyly in the same individual is a rare condition [10-13].

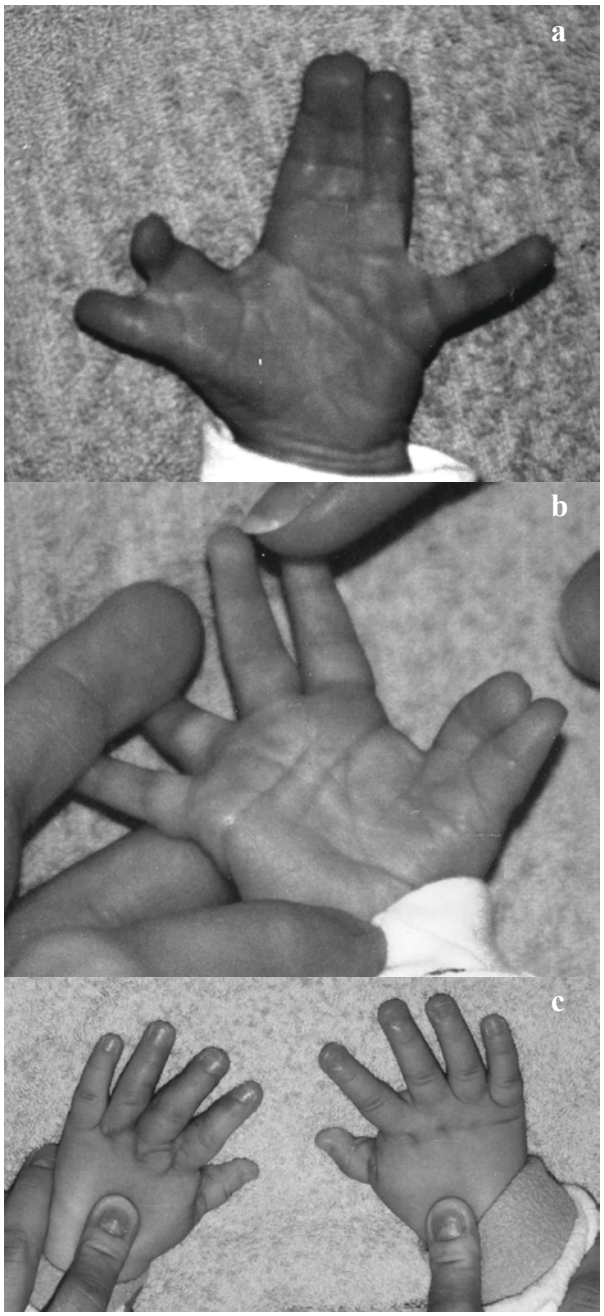
CASE PRESENTATION

A male infant born in 2000 presented polydactyly and syndactyly of hands and feet. The physical and X-ray examinations showed the extra thumbs well developed – preaxial polydactyly type A of both hands and syndactyly of the II-IV digits of the left hand (*Fig. 1A* and *1B*). On both feet there were polydactyly of the big toes, on the left side type A with syndactyly type I and on the right side in the first and second toe type B polydactyly with syndactyly type I (*Fig. 2A)*. He was operated in the first and second year of life. The accessory fingers were removed, syndactyly was operated with using zigzag method. The final scars are minimal, and functionally and cosmetically good results are obtained (*Fig. 1C* and *2B*). A genetic history of patient reported a presence of hereditary deformations of digits in eleven members of family, in three generations.

DISCUSSION

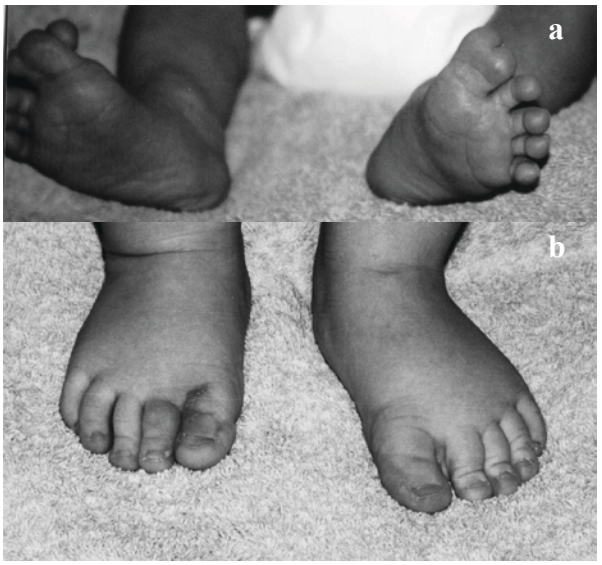
Syndactyly is a congenital deformity, with an incidence of approximately two or three per 10,000 live births. Inheritable syndactyism is associated with genetic defects on the second

Figure 1. **Hands.**
A) Left hand before the operation. Note the preaxial polydactyly type A and syndactyly of the II-IV digits.
B) Right hand before the operation. Note the preaxial polydactyly type A.
C) Hands after the operation.



chromosome. Polydactyly occurs in similar approximately as syndactyly but it is ten times more frequent in Blacks than in Whites [2,3,7,9,10]. Preaxial polydactyly of the feet and hands and both types of polydactyly –likes in this case- in the same individual is a very rare. Polydactyly occurs both in a sporadic form and in a hereditary form. Two different clinical pictures, and two different underlying dominant genes have

Figure 2. **Feet.**
A) Feet before the operation. Polydactyly of the big toe, on the left side type A with syndactyly type I. On the right side polydactyly type B with syndactyly type I between I and II fingers.
B) Feet after the operation.



been postulated [2,3,5,12,14]. In polydactyle type A a high penetrance determined by a single gene has been suggested. The genetics of type B seem more complicated and two dominant genes have been suggested. Venturo et al. [14] postulated that the case where both A and B types are in the same subjects depends on a single dominant gene. Different clinical pictures (A or B types) may be attributed to the influence of non genetic factors for example prenatal smoking [15,16], maternal diet [17] or exposure to ethanol [18]. Teenage pregnancy increases the risks of congenital limbs' deformities too[19]. Polydactyly and syndactyly occur alone or as a part of a syndrome. It often exists with syndromes of Gorlin [20], Klippel-Trenaunay [21], Curry –Jones [22], Carpenter [23], Mohr-Clausen [24], Apert, Klippel-Feila, Grieg [8,25], Merckel-Gruber [26,27], Smith-Lemli-Opitz [28], Barden-Biedl [29]. Classification and characterization of the hand malformation can help to narrow the differential diagnosis in spite of fact that some abnormalities are highly suggestive of a specific diagnosis [1].Our case presents isolated anomalies without any others changes in the newborns body but genetic history of patient showed a hereditary form of the anomalies. Deformities were observed in three generations in a mixed forms- type A and B, hands and feet. It suggests that the dominant mutant gene may express itself as the type A or B polydactyly. The diference in expression being due to modifiers. The similar situations were observed by Ventruto and al. [14] and Dukic and al. [30]. In our case the penetrance of the gene of polydactyly is higher in successive generation.

CONCLUSION

The prognosis for the patient seems to be good. Surgical correction is satisfying - the final scars are minimal, functional and cosmetical results are very good but the patients has a genetic load of presence of fingers' abnormalities in following generations.

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