Available Online: http://irjs.info/



A case of partial anonychia with a review of literature

Devendra D. Jalde*, Satyajit A. Godhi and Shashi M. Uppin

Department of General Surgery, KLE University's J.N.Medical College, Belgaum -590010. Karnataka, India

Abstract

Anonychia (absence of the nails) from birth is a rare congenital anomaly. It may occur as an isolated sign, which is very rare or be accompanied by other defects of the digits and other structures. Of these abnormalities, the most frequent are some hypoplastic nails, absence of distal phalanges etc, this is incomplete anonychia. The mode of inheritance of most of these disorders has not yet been established with certainty. Very few cases of anonychia with the above mentioned disorders have been reported but this is probably the first report of a case of anonychia along with congenital absence of umbilicus.

Keywords: Partial anonychia, congenital absence of umbilicus, Surgery, Diagnosis.

INTRODUCTION

Congenital anonychia or absence of nails since birth is considered to be rare. Very few cases have been reported in the past. Congenital anonychia without any other coexisting major anomaly (simple anonychia) is extremely rare. Congenital anonychia is always found associated with other abnormalities such as hypoplastic nails or absence of distal phalanges. Most cases of anonychia occur as part of a syndrome. There have been no previous reported cases of congenital anonychia with congenital absence of umbilicus. The first instance of such an occurrence is presented here.

CASE REPORT

A 37 year male patient presented to surgery department with complaints of ulcer over medial aspect of left leg, the patient was admitted. The patient gives history of a similar lesion over the same part which was operated 3 years ago (with excision and split thickness skin grafting). The present ulcer was arising from the same previous scar tissue. On further questioning, it was also noted that the patient had complete absence of nails in his lower limbs and partial growth of nails in the upper limb since birth. No history of similar complaints in any of his family members.

On examination

He was noted to have complete absence of nails in the lower limb with partial absence of nails in the upper limbs. Umbilicus was absent since birth. He was also noted to have lid lag of the left eye .A ulcer of 5x3 cms with everted edges and bleeds on touch. Old split thickness skin graft seen over the same area. Inguinal lymphadenopathy present.

Received: Feb 08, 2012; Revised: March: 15, 2012; Accepted: April 10, 2012.

*Corresponding Author Dr. Devendra D. Jalde Department of General Surgery, KLE University's J.N.Medical College, Belgaum -590010. Karnataka, India

Tel: +91-9986402123; Fax: +91-831 2470732

Email: dev_dj2000@yahoo.com



Fig 1. Lower limb Showing absence of nails



Fig 2. Upper limb Showing partial absence of nails



Fig 3. Skin graft placed over the medial aspect of right ankle joint and foot

28 Devendra D. Jalde



Fig 4. Healed skin graft

DISCUSSION

Anonychia (absence of nails) is a very rare congenital or acquired anomaly. Congenital anonychia has been described in association with a wide variety of other congenital anomalies. Of these abnormalities, the most frequent are some hypoplastic nails, this is incomplete anonychia. Simple anonychia, meaning congenital absence of the nails without any other coexisting major congenital anomaly, is an extremely rare variety of this condition. This is mostly due to autosomal recessive inheritance. A case with congenital but non-inherited absence of all toe nails and partial finger nails together with the congenital absence of umbilicus, without any other major structural anomaly is reported.

The term 'anonychia' describes the absence of fingernails and/or toenails $^1.$ It may occur as a single feature or as a part of a syndrome. Non-syndromal anonychia has been reported either in a partial or total form. The partial form usually involves the thumbs only, and it is inherited as an autosomal-dominant Trait $^2.$ Otherwise, anonychia represents an autosomal-recessive trait when it involves the second, third and fourth digits $^3.$

Some of the common causes for acquired anomaly are ichthyosis, severe infection, severe allergic contact dermatitis, self-inflicted trauma, raynaud phenomena, lichen planus, epidermolysis

bullosa or severe exfoliative diseases.

Anonychia is the absence of nails, a rare anomaly, which may be the result of a congenital ectodermal defect ⁴⁻⁶, ICD-10 L60.8, Q84.3, 1CD-9 703.8, 757.5

REFERENCES

- [1] Bergmann C, Senderek J, Anhuf D, Thiel C T, Ekici A B, Poblete-Gutierrez P, van Steensel M, Seelow D, Nurnberg G, Schild H H, Nurnberg P, Reis A, Frank J, Zerres K. 2006. Mutations in the gene encoding the Wnt-signaling component R-spondin 4 (RSPO4) cause autosomal recessive anonychia. Am. J. Hum. Genet. 79: 1105-1109.
- [2] Blaydon D C, Ishii Y, O'Toole EA, Unsworth HC, Teh MT, Ruschendorf F, Sinclair C, Hopsu-Havu VK, Tidman N, Moss C, Watson R, de Berker D, Wajid M, Christiano AM, Kelsell DP. 2006. The gene encoding R-spondin 4 (RSPO4), a secreted protein implicated in Wnt signaling, is mutated in inherited anonychia. Nat Genet.; 38(11):1245-7.
- [3] Diana C Blaydon, Yoshiyuki Ishii, Edel A O'Toole, Harriet C Unsworth, Muy-Teck Teh, Franz Ru schendorf, Claire Sinclair, Va ino K Hopsu-Havu, Nicholas Tidman, Celia Moss, Rosemarie Watson, David de Berker, Muhammad Wajid, Angela M Christiano & David P Kelsell. 2006. The gene encoding R-spondin 4 (RSPO4), a secreted protein implicated in Wnt signaling, is mutated in inherited anonychia. Nature Genet. 38: 1245-1247.
- [4] Hopsu-Havu V K, Jansen C T. 1973. Anonychia congenita. *Arch. Derm.* 107: 752-753.
- [5] Littman A, Levin S. 1964. Anonychia as a recessive autosomal trait in man. J. Invest. Derm. 42: 177-178.
- [6] Mahloudji M, Amidi M. 1971.Simple anonychia. Further evidence for autosomal recessive inheritance. J. Med. Genet. 8: 478-480.