

AMNIOTIC BANDS WITH INFANTILE DIGITAL FIBROMATOSIS

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Hariharasubramony Ambika¹, Chankramath Sujatha¹,
Srinivasiah Santhosh²

¹*Department of Dermatology, M.V.J. Medical College & Research Hospital,
Hoskote, Bangalore, India*

²*Department of Pediatrics, M.V.J. Medical College & Research Hospital,
Hoskote, Bangalore, India*

Corresponding author: Dr. Hariharasubramony Ambika ambs120269@yahoo.com

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A Newborn baby presented with deformed digits of both hands since birth. Baby was born as full term normal baby of weight 2.2kg to non consanguineous parents. No history of any medications taken or infections during antenatal period. Mother's. HIV and VDRL tests were non-reactive. There was no family history of similar illness. On examination child was healthy and alert. Vitals were WNL. Both hands showed multiple constriction bands over fingers with loss of distal phalanx of some digits. Portion of digit distal to bands showed diffuse swelling. There was a firm nodule of about 5mm diameter was present on right ring finger and left index finger. No other cutaneous or systemic abnormalities were observed. X-ray and Ultrasound scan of digital nodules revealed fibrous nature of digital nodule. Biopsy or FNAC of nodule could not be done due to lack of consent from parents. Child was referred to orthopaedic surgeon for release of adhesion bands and child is under regular follow up for nodular lesion which was diagnosed as Infantile Digital Fibromatosis clinically and confirmed by USS.

Amniotic bands are congenital constriction bands which occur due to rupture of amniotic membrane which happens usually before 12 weeks of gestation [1]. Small strands of amnion encircle developing structures commonly digits causing constriction bands (pseudoainhum) pseudosyndactyly, auto amputation or if occlusion is partial leading to distal lymphoedema [2]. Large bands can cause decreased foetal movements which can be detected in utero by USS [3]. Bands in ankle joint can cause club foot or if it is in trunk can cause scoliosis. Facial clefts in association with amniotic bands are reported [4]. No two affected babies will have exactly the same features and there is no single feature

that occurs consistently in all cases. Examination of placenta may reveal strands of amnion rolled up at the base of placenta. Results of amnion rupture are external so no internal anomalies are associated. Constriction bands can also be seen due to external forces like hair or thread. It can occur secondary to diseases like palmoplantarkeratoderma or after infection, trauma and some times seen associated with Michelin tyre baby [5]. Auto amputation occurring in utero has to be differentiated from hypoplasia aplasia and acromelia. Amniotic adhesions are also seen with limb body wall complex defects which is due to different pathomechanism.

Infantile Digital Fibromatosis are rare cutaneous juvenile Fibromatosis [6]. They are also called as Rey tumours as they are first described by Rey in 1965. They are usually present at birth as nodules over 3rd to 5th digits or may occur after birth. Histopathologically show paranuclear eosinophilic inclusion bodies inside interlacing bundles of myofibroblasts [7]. Inclusion bodies stain red with masson's trichrome stain. Electron microscopically these are actin filaments. Infantile digital Fibromatosis has to be differentiated from conventional fibromatosis. So biopsy is mandatory. Conservative treatment is recommended due to benign nature and spontaneous regression. However rapid growth and functional impairment may necessitate surgery. 60% shows recurrence after surgery. Intralesional steroid and bleomycin has been found effective [8].

Association of amniotic bands with infantile digital fibroma has not been reported in literature to our knowledge.



Figure 1. Infantile digital fibromatosis



Figure 2. Infantile digital fibromatosis

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Hariharasubramony Ambika, Chankramath Sujatha, Srinivasiah Santhosh

Dr. Daisuke Tsuruta PhD, Dr. Teruki Dainichi, Dr. Takahiro Hamada, Dr. Norito Ishii,
Dr. Takashi Hashimoto

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Amniotic bands are uncommon conditions that may lead to malformations and fetal-infant death [1]. Amniotic bands are congenital constriction bands from the outer membrane surface into the amniotic cavity, that occur when the amniotic membrane ruptures [2]. As baby develops, amniotic bands can trap extremities and may cause immobilization, constriction or even amputation of the structure [2]. It is not so common for Dermatologists. Even Fitzpatrick's textbook does not have section for it. However, as amniotic bands compress skin directly, dermatologist should have the knowledge of it. In this report, Dr. Ambika and his colleagues showed a complication possibly caused by amniotic bands [2]. They showed that infantile digital fibromatosis was associated with amniotic bands in a newborn baby [2].

Infantile digital fibromatosis is characterized clinically by asymptomatic, flesh-colored and firm nodules affected on the fingers and toes in infants [3]. It may be present at birth. The most affected sites on fingers are the third to fifth digits [3]. It is histopathologically characterized by poorly circumscribed, interlacing bundles of myofibroblasts, in which eosinophilic, Masson trichrome stain-positive paranuclear inclusion bodies are observed [2,3]. These inclusion bodies are the hallmark of infantile digital fibromatosis, differentiating this from other conventional fibromatosis [2,3].

The unfortunate thing is that the patient did not consent to perform skin biopsy of the lesion. Therefore, as mandatory (the authors used this word in the article) biopsy for the diagnosis of infantile digital fibromatosis was not performed, it is possible that the diagnosis may have been wrong.

However, we agree the clinical diagnosis of infantile digital fibromatosis because of the typical and convincing clinical photos they showed in the article and the result of ultrasound scan.

The pathogenesis of infantile digital fibromatosis is unknown. However, it has been suggested the roles of transforming growth factor- β 1 mediated differentiation of myofibroblasts from fibroblasts and of bone morphogenetic protein-mediated apoptosis [4,5]. Therefore, it is tempting to speculate that amniotic bands may enhance the expression of these growth factors in the fetus.

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Department of Dermatology, Kurume University School of Medicine, and Kurume University Institute of Cutaneous Cell Biology, Kurume, Fukuoka, Japan

Correspondence:

Dr. Takashi Hashimoto,

E-mail: hashmot@med.kurume-u.ac.jp