RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 7 | July 2015 | ISSN - 2249-555X

A Case of Congenital Amniotic Band Syndrome Involving All the Four Limbs With Severe Anemia With Thrombocytosis: A Rare Occurrence in an Elderly Adult.

KEYWORDS: Congenital Amniotic Band Syndrome, Streeter's dysplasia, Congenital Constriction Bands.

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ABSTRACT *A 55 yr old female patient presented with generalised weakness, easy fatiguibility and loss of appetite since one month and was also having congenital deformity of all the four limbs. After investigating and thorough examination a diagnosis of congenital amniotic band syndrome, severe anemia and thrombocytosis was made.*

Introduction:
Congenital amniotic band syndrome is a rare congenital abnormality with multiple disfiguring and disabling manifestations. A wide variety of clinical deformities are encountered, ranging from simple ring constrictions and minor digital defects to major craniofacial and visceral defects[1].We encountered a case of congenital amniotic band syndrome along with anemia in a fifty five years old woman.

Case history:
A 55 year old female patient presented with generalised weakness, easy fatigability and loss of appetite since one month and was also having congenital deformities of all the four limbs. She was able to walk and was able to do all routine daily activities. On examination she had pallor and congenital limb deformity as shown in Figure 1 and Figure 2. Rest of the examination was normal.

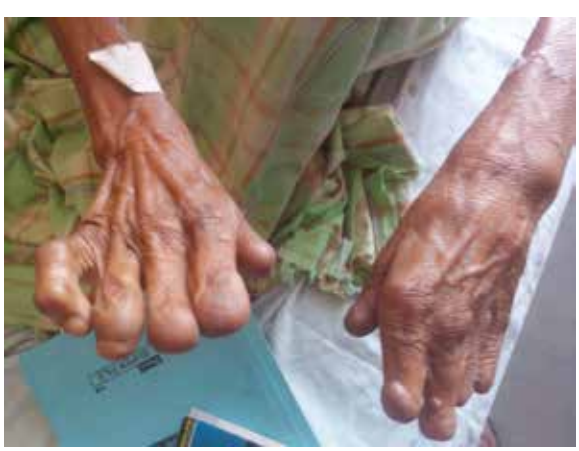

Figure 1

Figure 2

On investigation her haemoglobin was 5.1 gm%, TLC was 14500/mm3 and platelet count was 785000/mm3. Peripheral smear showed microcytic hypochromic anemia with anisopoikilocytosis, TLC was raised and platelets were scattered and adequate. Bone marrow aspiration examination was suggestive of erythroid hyperplasia with normoblastic maturation and megakaryocytosis. Liver function and renal function tests, abdominal sonography, Chest X ray P/A view were within normal limits. Orthopaedic, Surgical and Ophthalmic evaluation was obtained. After ruling out oth-

Volume : 5 | Issue : 7 | July 2015 | ISSN - 2249-555X

er causes, we diagnosed her to have a case of congenital amniotic band syndrome with anemia and thrombocytosis. We transfused her two bags of whole blood. She was not advised surgery by orthopaedic surgeons as she was able to do routine activities and was elderly. The patient also did not want to get surgical correction for cosmetic reasons. She was feeling better after blood transfusions so she was discharged and asked to return for follow up.

Conclusion:
A 55 year old female patient presented with generalised weakness, easy fatigability and loss of appetite since one month and was also having congenital deformities of all the four limbs. After investigating and thorough examination a diagnosis of congenital amniotic band syndrome, severe anemia and thrombocytosis was made.

Discussion:
A congenital constriction band is defined as a disorder present in a newborn infant, in which constriction rings or bands cause soft tissue depressions, encircle digits, extremities or limbs and rarely in the neck, thorax, and abdomen [1]. We encountered a case of constriction bands who was 55 years of age. Hence this report.

It is a rare, sporadic condition with an incidence of 1: 11,200 births and no sex predilection. Antenatal diagnosis of this syndrome is difficult because the amniotic bands may be very thin and difficult to identify on ultrasound, unless a thorough search is performed[4]

The condition is also known as ADAM complex, amniotic band sequence, Streeter's dysplasia, congenital constriction bands, and pseudoainhum. They are associated mostly with other congenital anomalies such as syndactyly, clubfoot, cleft palate, and cleft lip and can also present with severe craniofacial and visceral deformities.[2]

Familial occurrence is very rare, and there is no known genetic predisposition and/or gender difference. The pathogenesis is unknown; however, a number of theories are postlated[1]. The exogenous theory, which proposes the early partial rupture of the amniotic sac leading to fibrous bands; these fibrous bands float in the amniotic fluid and can encircle and entrap a part of the fetus. These act as constricting bands as the fetus grows, causing reduced blood circulation, which can lead to autoamputation of a digit or limb in utero. In some cases, it leads to necrosis requiring surgical amputation following birth.[2] Amniotic bands are composed of either acellular fibrous tissue or fibrous tissue containing fibroblasts, covered by squamous cells . The depth of a lesion varies from a shallow groove to a deep gutter and usually extends only up to the first fascial layer. Histology shows a grossly normal epidermis over a thickened dermis with collagen bundles and elastic fibers and large amounts of hyalinized connective tissue[1] George Linus Streeter, the director of embryology at the Carnegie Institute, believed, wrote, and taught that the etiology of the condition was primarily defective germ plasm. Because of his work many physicians continue to refer to this condition as Streeter's dysplasia[3] Torpin has cited a genetic aetiology caused by germ plasm defects, calling this syndrome "fetal focal dysplasia". However, this has not been widely accepted as it fails to explain the tremendous variety and asymmetry of lesions involving structures formed at different times in uterine life and derived from different germ layers.[4]

Amniotic disruption theory by Torpin et al is the most widely accepted theory. This theory suggests that amniotic band syndrome occurs when amnion ruptures before 12 weeks of gestation resulting in the chorionic side of amnion emanating numerous mesoblastic fibrous strings which entrap fetal parts. As the fetus develops, the amniotic bands can trap extremities causing immobilization, constriction or amputation of structure. Early insult results in facial clefts and brain defects, while late insult (after 45 days gestation) results in limb involvement without facial clefting or CNS involvement.[5]

When bands are superficial, only mild skin indentation occurs. Deeper, tighter bands may produce profound compression with obstruction of both venous and more dramatically lymphatic drainage. Deep compression may result in considerable edema in the digit or limb distal to the band.[3]

The treatment of the circular constriction band can be accomplished by the removal of the fibrotic tissue with 1 to 2 mm of intact skin and by adding z-plasties or w-plasties to avoid further constriction of the scar. Subcutaneous tissue deficiency under the constriction ring can be corrected with rectangular plasty techniques and by using turnover dermofat flaps. If the constriction is around the digits or extremities, urgent surgical treatment may be necessary because of vascular compromise. Otherwise, the treatment is usually performed for cosmetic reasons, and a staged correction may ensure the adequacy of vascularity to the residual limb or digit.[1]

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