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CASE REPORT

Late Presentation Of Amniotic Band Syndrome A Case Report

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ABSTRACT

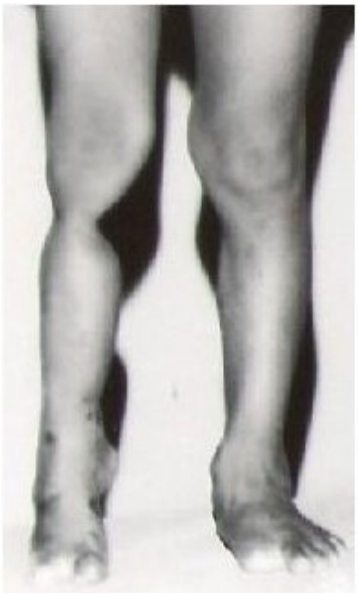
Amniotic band syndrome is a term used interchangeably to Streeter's dysplasia and refers to a continuous spectrum of manifestations occurring due to intrauterine rupture of amnion. The disease may be characterised by simple soft tissue constriction bands to amputation of digits or more severely of the whole limb due to dysplastic vasculature. Due to digital amputation the condition is also known as pseudo-ainhum. This is a rare condition. Therefore, we report a case wherein at the age of five years there are constriction bands in the right leg while amputation was noted in toes and in the right index and middle finger.

Introduction

Streeter's dysplasia, which is also known as amniotic band syndrome, is a rare abnormality that manifests itself at birth in the form of ring-like constrictive bands either in the upper or in the lower limbs and occasionally in the trunk [1]. These annular defects occur in varying degrees of severity, ranging from superficial circumferential grooves in the skin to amputation of a part or the whole of a limb. A vascular insult occurring very early in the gestation prior to 26 days is considered to be the causative factor. Associated defects with amniotic band syndrome include hydranencephaly, porencephaly, craniofacial abnormalities, and spinal dysraphism [2]. We report a case of amniotic band syndrome with multiple bands involving both the upper and lower limbs with varying degrees of tissue loss distal to the constrictive bands.

Case report

A five-year-old male child presented with the chief complaints of multiple constrictive rings in both upper and lower limbs. The child was the first born of his parents and delivered full term at home through a normal vaginal birth. The constrictive rings noticed since birth were present on the right lower limb (Table/Fig 1), toes (Table/Fig 2), and fingers of right upper extremity (Table/Fig 3).



Table/Fig 1 Frontal view of both legs showing a deep costriction ring in the right leg with trophic changes distally

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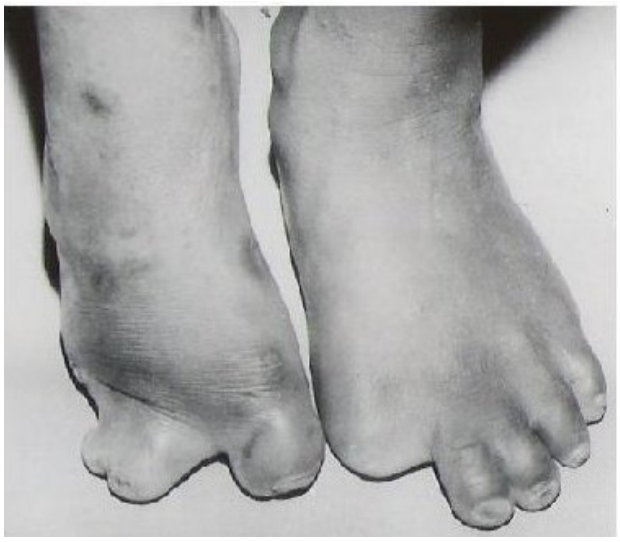
These rings were shallow at birth but gradually became deeper. Local examination revealed a deep constrictive circumferential groove at these sites. To begin with, there were no neuro-vascular symptoms in the parts distal to the bands. Subsequently, from the age of two-years, he complained of a tingling sensation in the toes and fingers. This was followed by amputation of tissues distal to constrictive bands by the age of 4½ years.

The psychomotor milestones of the child were normal. No contributory family history was present. Maternal obstetric history was not suggestive of any antenatal insult to the foetus. A skeletal survey revealed a deep groove at the lower ends of the tibia and fibula in the right leg.

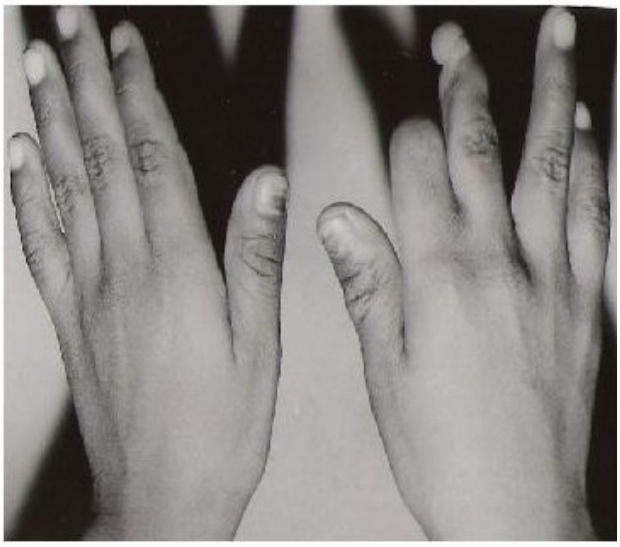
The groove was confined to the skin and the subcutaneous tissues, the bones being normal in this area. There was loss of both soft tissue and bone in the digits of the right hand and both the feet. The fibrous band in the right leg was excised and Z-plasty done. Subsequent development of the leg and foot was normal. For the amputated digits, reconstructive surgery was offered. The parents however, declined the same due to financial constraints.



Table/Fig 2. Frontal view of both feet showing amputations of multiple digits.

Discussion

The prevalence of amniotic band syndrome among live births is estimated to be around 7.7:10,000 [3]. Among abortuses it may be as high as 178:10,000 [5]. It affects males and females in the same proportion. Most of the cases are sporadic, with no recurrence in siblings or children of affected adults [4].



Table/Fig 3: Frontal view of both hands showing amputation of digits as well as trophic changes.

Pillay and Hesketh [6] reported the largest series of 40 cases from Singapore and concluded that the disease is more common in the Malaya people of the Far East. In the lower extremities, the incidence is a little higher than the upper extremity. The usual sites for these annular grooves are the distal parts of the limbs. These grooves typically occur in fingers, toes, forearms, and legs. Occasionally, these are found above the knees and elbows. A rarer site is the trunk. These rings may be very shallow involving the skin and subcutaneous tissues only or they may be deeper and involve the deep fascia. involving [1].

The condition is idiopathic. Maternal trauma, oophorectomy during pregnancy [7], intrauterine contraceptive device [8] and amniocentesis [9] and familial incidence of connective tissue disorders (Ehler-Danlos syndrome) [10] are some of the implicated etiopathological factors. Lockwood C et al suggested the role of a teratogenic insult as one of the factors causing the disorder [11]. The cause of the malformations in the amniotic band syndrome remains controversial. These may be due to focal developmental errors in the formation of limb connective tissue. According to Torpin et al, the rupture of the amnion without the rupture of the chorion leads to transient oligohydramnios due to loss of amniotic fluid through the initially

permeable chorion [12]. The foetus passes from the amniotic to the extra embryonic coelom through the defect and comes in contact with 'sticky' mesoderm on the chorionic surface of the amnion. This leads to entanglement of the foetal parts and skin abrasions. Entanglement of the foetal parts causes constriction rings and amputations, whereas skin abrasions can lead to disruption defects, such as cephaloceles. Further, swallowing of the bands will cause asymmetric clefts on the face [12].

The most common finding in the amniotic band syndrome is the constriction rings of the fingers and toes [13]. It occurs in 77 percent of foetuses with multiple anomalies [5]. The association of abnormalities, such as clubfeet, anencephaly, cleft lip, cleft palate, cleft face, rib clefting, gastroschisis, omphalocele, bladder exstrophy, and imperforate anus, should be regarded as strong evidence and should arouse the suspicion of the amniotic band syndrome. The visualization of amniotic bands attaching to a foetus with restriction of motion on prenetal ultrasonography is diagnostic of the condition. This precludes the need for foetal karyotype [5,14]. The visualisation of amniotic bands or sheets in the absence of foetal deformities should by no means lead to the diagnosis of the amniotic band syndrome since several types of membranes, such as folds of amnion and extra-amniotic pregnancies [15] may be seen in normal pregnancies [14].

The differential diagnostic considerations include ammiotic folds, short umbilical cord syndrome and extra-amniotic pregnancy. Ammiotic folds which are recognized by prenatal ultrasonography as reflecting membranes floating freely in the amniotic fluid. They have been reported in patients who had instrumentation of the uterine cavity resulting in intrauterine scars or adhesions. The foetus is morphologically normal. In short umbilical cord syndrome, also known as limb-body wall complex or body stalk anomaly, there is a complex set of disruptive abnormalities having in common the failure of closure of the ventral body wall. Although some of the features are similar to those of the amniotic band syndrome, there are distinctive characteristics: marked scoliosis, evisceration of abdominal contents into the extraembryonic coelom, and a shortened umbilical cord [16]. Limb amputations are not typically found in this syndrome, which is predominently limited to the abdominal wall. Extra-amniotic pregnancy-amniotic pregnancy is a rare condition that is explained by a mechanism similar to the exogenous theory of the amniotic band syndrome: a rupture of the amnion, with development of the foetus in the extra embryonic coelom.

Associated anomalies decide the ultimate prognosis. This can be quite good for infants with minor constriction rings and lymphedema of the digits while children with amputations of the limbs may require either reconstructive or plastic surgery and prosthesis. Good results have been reported in the literature by using Ombredanne's two-stage operation or serial excision and repair with Z-plasties [17]. There is normal life expectancy for these cases. The syndrome is lethal for the severe forms with multiple associated anomalies [4],[5].

Photographs: All Photographs displayed in this article were obtained after informed consent by the parents of the child.

Conflict of Interest: None declared

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