

UNUSUAL PRESENTATION OF AMNIOTIC BAND SYNDROME

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Introduction

Amniotic band syndrome (ABS) is a sporadic condition¹; the term is used to describe a complex disorder characterized by constricting rings, acrosyndactyly and often amputations of the extremities of neonates. It was recognized as early as 300 BC by Hippocrates² who suggested that extrinsic pressure from a ruptured amniotic membrane result in band formation or digital amputation.

In 1930, George Streeter³ introduced his theory of germ plasm defect as one reasonable etiology. He proposed that a disruptive event occurs during blastogenesis, leading to an intrinsic germ plasm defect. This causes the soft tissue to slough. External healing of the slough leads to the constricting rings and the resultant localized developmental defect. His theory was well accepted at that time because of the associated distant anomalies occurred far from the site of the constriction bands, he called his theory "Streeter dysplasia". In 1960, Patterson⁴ study the histology of normal skin creases to show their similarity to constriction bands. He hypothesized that the same lack of mesodermal development occur in the area of the band, thereby making the bands simply abnormal creases⁴.

Richard Torpin⁵, in 1965, examined many placentae and infants with the disorder. Torpin reintroduced the extrinsic theory (Hippocrates theory). In

his study of fetuses and placentae, he noted the lack of a complete amniotic lining in the placenta of neonates with ABS. Strands of amnion were also visible around constricting rings of the digits, and binding strands were visible at the tips of limbs with acrosyndactyly. He proposed that intrauterine trauma led to premature rupture of the membranes, and strands of residual membranes could encircle the digits or might even be swallowed. He proposed that maternal trauma led to rupture of the amniotic membrane, which then formed into strands. These encircling strands cause extrinsic compression on the head or limb, leading to the formation of constricting bands, vascular occlusion, and eventually, amputations. Currently, this is the most widely accepted hypothesis; therefore, this disorder would be more accurately termed ABS⁵.

The estimated frequency of ABS world wide is 1 case per 10,000-15,000 population². In Australia the incidence rise to 2.03 per 10,000 births⁶.

Clinical features

Children born with ABS are usually full term or a few weeks premature; in most cases, the pregnancies were uncomplicated. In Light and Ogden's series⁷, the average gestation was 37.5 weeks, with an average birth weight of 3.0 kg. The most common clinical findings in ABS appear on the extremities and include amputation followed by constriction bands and then acrosyndactyly².

On the extremities, the distal portion is most often involved; especially the longer central digits of the hand (middle, long, and index fingers)⁷. The hands are affected in almost 90% of cases, with involvement of the thumb or small finger², presumably because of their shorter lengths. The same rule holds true for the feet, where constriction bands most commonly involve the halux⁷. Mild band pressure causes only indentations at the base of the phalanx, usually distal to the metacarpophalangeal joints.

Constriction is due to the maceration of the indented skin, then healing by scar tissue formation². If the compression from the band is severe, lymphatic and vascular compromise may ensue, and the child presents at birth with a swollen engorged digit or limb that may require immediate surgical release. More commonly, the digit has been amputated in utero.

Depending on the severity of the constriction, the defect could be as minimal as a merely cosmetic band, deeper bands may cause lymphatic obstruction leading to edema and vascular compromise that necessitates immediate release. Pressure from the bands may potentially cause abnormalities distal to the constriction, such as hemihypertrophy, anterolateral bowing, pseudarthrosis, leg-length discrepancy, and resistant teratologic clubfeet².

Case report

One month old female child was referred to the Plastic Unit in the Teaching Hospital in Basrah, with huge swelling of the lower half of left leg and the foot and amputation of proximal part of the right Index.

The child is the first of a consanguineous marriage. During pregnancy the 19 year old mother had a transient hypertension during first trimester, but the rest of the pregnancy passed with no complication. Before the time of

delivery the midwife suspects breech presentation so the mother was referred to the Basrah Maternity Hospital, but she had normal delivery of a full term otherwise normal baby girl.

On examination, large lymphoedema swelling of lower part of left leg and foot with complete loss of the outline of the foot and toes (Fig.1&2), the toes also show some bands reconstruction and digits amputation. The swelling is covered by thin darker skin.

Fig.1



Fig.2



The swelling at proximal part of the foot measures 17 cm in width, and too heavy for the child to move it. Careful inspection of the swelling show two constricting bands at lower leg, the proximal one form a complete circle but the distal band which is at ankle area is not circumferential, nevertheless, both were deep enough to reach the deep

fascia (Fig.2). There is another constricting band on the right leg but with no swelling. The index finger of the right hand had an intrauterine amputation through middle phalanx, (Fig.3).



At the age of three months the child was taken to theater, the upper constricting band was excised totally till the deep fascia; about 200 milliliters of clear lymph were seeping from the distal end of the wound, also fresh blood were coming too from both ends of the wound. The healthy tissue was approximated with two large Z plasty flaps which include skin and subcutaneous tissue placed at both sides of the leg (Fig.4).



The lower band was excised as well and closed with Z plasty. On the right side the band was left alone because there was no distal swelling.

Immediate post operative period was smooth, and the child was discharged on second post operative day. The sutures were removed after ten days with good wound healing (Fig.5).



The child was reviewed at age of six months; the swelling was significantly reduced with plenty of redundant skin which needs to be excised and trimmed later (Fig.6&7). She can move her leg easily but the toes show a lot of toe amputation at different levels.



Discussion

Amniotic band syndrome is a rare entity wide spectrum of associated congenital anomalies, usually affect young mother in her first pregnancy⁸. More commonly affect upper limb than lower limb⁷, The extremities were most often affected distally⁹. Intrauterine amputation of the digits affect the index, middle and ring fingers more commonly. In foot, it's the halux which is more prone for amputation⁷.

In the presented case, the left leg is the affected limb with this enormous unusual swelling this is a rare presentation of ABS, but index is the usual digit affected as in Light & Ogden series³.

Surgical treatment is urgent when there

is compression of vascular bundle². In this case the vascularity was good with pink color warm sole, so there was no hurry in release of the constricting band.

Some authors do the excision of the constricting band in two stages with or without z plasty^{2,8} to avoid compromising the blood supply of the limb, while others advocate one stage excision of the whole band with reconstruction with or without z plasty^{10,11}.

Nothing can be done for the index now, functionally its good. In the future, cosmetic prosthesis can be used. Children who had intrauterine amputation are generally well adapted to their physical limitation².

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