Amniotic bands
A case report
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Summary
The aetiology of amniotic bands causing intra-uterine annular constrictions of the fetus is discussed and a case report is presented. Cautious use of amniocentesis in the first trimester appears to be advisable.


Annular constrictions and intra-uterine amputations are believed to be the result either of an embryological development defect of the germ disc, an endogenous cause,1 or a mechanical constriction band from early rupture of the amnion, an exogenous cause.2

A third theory that they may be due to an exogenous superimposed disease process of unknown aetiology, as suggested by Stocks and Stocks's original theory, is discussed and a report of a case in which the aetiology was possibly hereditary is presented.

The importance of a detailed history as well as histological examination of the placenta, the cord, the attachment sites of the bands and the sites of embryological abnormalities of the fetus should be stressed in gathering information on this condition.

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The monstrosity is contrary to nature, not contrary to nature taken absolutely but contrary to the most usual course of nature. Nothing in fact can be produced contrary to that nature, which is both eternal and essential.

Aristotle, 4th century BC

Radiographs of any affected limbs should also be taken and congenital abnormalities should be looked for. Attention is drawn to the increased use of amniocentesis in early pregnancy; attending staff should be alerted to these anomalies and to the possibility that this procedure may precipitate amniotic damage.1

Chemke et al.5 give an incidence for annular constrictions of 1 in 5000 to 1 in 10 000 pregnancies.6,7

Case report
An infant was born prematurely on 31 January 1979. Before delivery the gestational age was estimated as 32 weeks by means of ultrasonography, and no fetal abnormalities were detected. The mother was a 23-year-old White woman, para 0, gravida 1; her last menstrual period had commenced on 10 June 1978 and the expected date of delivery was 17 March 1979. She was initially seen at 20 weeks' gestation and again at 24 weeks when ultrasonography was carried out because the patient was large for dates and had polyhydramnios; twins were suspected but only a single fetus was found. The mother, a teacher, gave a history of rheumatoid arthritis since childhood and had been exposed to acetylsalicylic acid and unknown homeopathic remedies in the first 12 weeks of her pregnancy. There was no family history of congenital abnormalities, rhesus disease, diabetes mellitus or hypertension in pregnancy on the maternal side. The father, a 25-year-old draughtsman, gave a history of club feet in his mother and cleft palate in a first cousin.

On admission the patient was in premature labour; it was attempted to stop labour with intravenous hexoprenaline (Ipradol) 50 mg/l, regulated to maintain the pulse rate at under 120/min, but a fetus with gross congenital abnormalities was delivered. The baby's Apgar score at birth was 8 but dropped to 2 after 5 minutes, and she died 6 hours later.

The upper lip and forehead showed deep facial fissures (Fig. 1). The upper and lower limbs were well formed, but some digits of the hands and feet had band constriction. There was a constriction of the right hallux and second and third toes of the left foot (Fig. 2), a constriction of the base of the first finger, and a missing index finger on the same hand. There was a deep
A diagnosis of placenta with amniotic bands was made, based on histological examination of the placenta and cord and the external appearance of the fetus. It should be stressed, however, that the histological appearance of the placenta alone is not sufficient in making a diagnosis; the site of the embryological defect of the fetus should also be subjected to histological examinations. 2

On examination the placenta weighed 310 g and measured approximately 13.5 cm in diameter and 2 cm in thickness. The site of the membrane opening was marginal and the umbilical cord was eccentric in position, 12 cm in length and 1.3 cm in diameter. A false knot was present 1 cm from the site of insertion. Two amniotic bands could be seen on the fetal surface, one being 5 cm and the other 1 cm in length. The maternal surface was intact. On placental section, microscopic abnormality could be seen. Microscopic examination showed the chorionic plate, with normal amnion and moderate meconium deposition. Amnion was present in the area of the bands and foci of amnion nodosum could be seen. The basal plate and vessels appeared normal. The fetal vili were mature with foci of villous immaturity, and mild cytotrophoblastic proliferation could be seen. The umbilical cord contained three blood vessels and was oedematous.

The parents were referred for genetic counselling and reassured that this condition was sporadic and unlikely to recur in subsequent pregnancies. In January 1980 the patient gave birth to a normal 2700 g live male infant. He measured 46 cm in length and was delivered by caesarean section for obstructed labour due to a large dermoid tumour arising from the left ovary, which was removed at the same time.

Discussion

The placenta, chorion and amnion are vital organs in the development of the fetus. Changes affecting placental tissue are often reflected in fetal growth and development. 3 There are very few primary diseases of the placenta that can be considered to relate directly to maldevelopment, namely amniotic bands, although there are secondary placental lesions which correlate with abnormalities such as fetal growth retardation, placental infarction, congenital syphilis, toxoplasmosis and viral infections.

A variety of amputations by amniotic bands has been documented but there is some doubt whether there is a causal relationship to other malformations, such as facial clefts. 4,5

The pathogenesis of amniotic bands is uncertain but a history of early prenatal bleeding may be associated with their presence. The amniotic sac does not become fully distended and applied to the chorion until the end of the first trimester. If rupture occurs before this, amniotic bands may form. 6

Congenital malformations, on the other hand, are common causes of neonatal death and although congenital constriction rings and intra-uterine amputations are seldom lethal, the role of the fetal membranes and the production of certain defects may explain deformities incompatible with extra-uterine life. 7,8

The first reported case of an intra-uterine amputation was by J. B. van Helmont (1577-1644). He mentioned several cases of limb defects in infants, which were then thought to be due to the sight of cripples by the pregnant mother. Besides the belief that these particular limb malformations could be produced by maternal impression, several other theories arose out of the confusion surrounding their production. These may be historically summarized as follows: (i) gangrene of the part; (ii) inflammatory exudates of the amnion producing 'bands' which encircle fetal parts; (iii) abnormal disposition of a portion of the amnion; (iv) maldevelopment of the amnion; (v) defective development of fetal germ plasma; (vi) penetration of the

constriction on the left middle finger over the distal interphalangeal joint (Fig. 3). The cause of the gross abnormality was investigated by means of histological examination of the placenta and fetal skin and heart blood chromosome analysis in case of a chromosomal abnormality. Subsequently the skin fibroblasts and heart blood revealed a normal female karyotype of 46XX. Autopsy was unfortunately not carried out and radiographs of the affected limbs were not taken.
amnion to various degrees by fetal extremities with consequent encirclement and constriction of the protruding part; and (vii) premature rupture of the amnion with consequent separation from the chorion leading to the development of chorionic or amniotic strings which encircle fetal parts.

From the above it can be seen that there has been a full swing of the pendulum from an exogenous mechanical origin of band constriction deformities to endogenous errors in fetal development and back again.7

Whether these band anomalies are endogenous defects within the embryo or exogenous forces acting in utero has been discussed by Streeter,1 Torpin2 and Stocks and Stocks.3

The hyalinized fibrous material of the constriction band was thought to arise deep within the involved fetal tissue and sometimes to originate before the 14th week of gestation as a local embryological defect.1 These endogenous deformities have been divided into genetic and teratogenic types.2 Genetic anomalies are derived from abnormal genes inherited from one or both parents, with a tendency to be transmitted to subsequent generations. Polydactyly, trisomy 21 and arrested or abnormal development of an extremity or digit are the malformations most often involved, but there may be other organ–system malformations, easily recognized in most cases.

The teratogenic malformations due to the effects of radiation, chemicals, drugs, specific toxins or viruses on the embryo at an early stage of development are often difficult to distinguish from the genetic unless there is an outbreak associated with the use of specific teratogenic agents, for example thalidomide, or with viral infections, e.g. rubella, but genetic teratogenic association with amniotic bands has not been proved.4,5

Torpin,6 the only investigator to study both the placenta and the membranes in the fresh state, believes that the amnion ruptures early in pregnancy with absorption of amniotic fluid and subsequent compression of the fetus. The chorion then thickens, resulting in retention of amniotic fluid, and the surface proliferation produces fibrous strings. When examined histologically these excised bands are not compatible with scar formation. Torpin states that they may relate to epithelial immaturity or to the biological activity of amniotic fluid resulting from immune tolerance. His observations on human fetuses have been substantiated by several well-controlled animal experiments.

Poswillo11 studied the effect of amniotic puncture before palate and lip fusion in rats; all the animals developed a cleft palate. In addition associated anomalies were noted, namely clubbing of the limbs, ring constrictions and amputations identical with those found in human fetuses with amniotic band malformations. Torpin12 believes that these exogenous constriction bands are amniogenic in origin. They have certain distinctions which should enable the clinician to distinguish them as such.

Absence of limbs or digits is usually multiple and unilateral but may be bilateral, asymmetrical and associated with constriction. According to Torpin,7 associated distal anomalies such as clubbing of the feet, lymphoedema and syndactyly produce a classic triad. There are usually no associated malformations of the internal organs; the placenta and membranes in the fresh specimens are sometimes normal. The remainder of the skeleton has no developmental abnormalities proximal to the level of the defect; any attempt at regeneration of the missing part is incompatible with an amniogenic origin. In exogenous syndactyly, fusion originates at the distal portion of the digit and may often be associated with a band of tissue encircling the involved member. In contrast, endogenous syndactyly of genetic or teratogenic origin is that in which the fusion includes the base of the digits.

Stocks and Stocks,7 in support of Streeter’s15 concept of an arrest in development before 14 weeks, favour the recognition of a secondary superimposed disease process of unknown aetiology such as infection, radiation or drugs causing focal secondary changes in developing tissues.

Conclusion

There are some families in which local limb defects are primarily hereditary, but it is conceivable that mechanical factors might result in local limb defects. Teratogenic and environmental factors causing secondary changes in developing fetal tissue specifically applicable to the first trimester may also be important in the aetiology of amniotic bands and local defects. Active movement of the fetus is also thought to be needed and is a necessary component of normal fetal growth. Medical personnel should be on the alert for malformations, especially with the increased use of amniocentesis and fetoscopy in early pregnancy. Close supervision and follow-up of patients who undergo this procedure may well be advised in view of the animal experiments of Poswillo11 in which all of the animals developed a cleft palate after amniocentesis.1,3,5 The pathogenesis of amniotic bands and the role of fetal movement remain uncertain, but an explanation of the deformities may come to light with closer observation of the affected fetal membranes and placenta and the increased use of real-time ultrasonography.

REFERENCES