Amniotic Band Disruption Sequence

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Introduction

A disruption of limb vasculature after the period of embryonic development can result in structural anomalies. This disruption can be due to tangled strands of amnion that encircle embryonic structures, placental emboli or death of a monozygotic twin [1]. Up to 50% of cases have other congenital anomalies including cleft lip, cleft palate, and clubfoot deformity [2]. Hand and finger anomalies occur in up to 80%. Other abnormalities found with amniotic band syndrome include clubhands, cleft lip and/or cleft palate, and haemangioma. The incidence varies from 1 in 10000 to 1 in 15000 live births.

Case Report

A term appropriate for gestational age female baby, weighing 2.5 kg was born to a booked immunized primigravida by normal delivery at our hospital. The mother and father were nonconsanguineous and were aged 24 and 26 years respectively. The pregnancy was complicated by vaginal bleed in the first trimester and severe abdominal pain during early second trimester for which she was given several medications including dicyclomine, ranitidine, metronidazole, iron and zinc preparations, norfloxacin, aminocaproic acid, uterine relaxants and antiemetics by various local practitioners. There was no history of trauma to abdomen or radiation exposure during antenatal period and she was non smoker and non alcoholic. The ultrasound examination during pregnancy did not reveal any anomaly. Postnatal examination of the placenta was normal and histopathology did not reveal any significant findings. The baby had an amputation defect of the right hand and a laterally bent left leg with calcaneovalgus deformity of the foot. There was also amputation of the digits of right foot (Figs. 1, 2). Examination of the respiratory system, cardiovascular system and abdomen was normal. The pelvis and the acetabular fossae appeared normal. There were no other skeletal defects present. Ultrasound examination of abdomen and cranium was normal. The baby was referred to orthopaedics department for

prosthetic device of the lower limb and was advised to return regularly for follow up.

Discussion

Amniotic band syndrome [3] is a set of multiple malformations due to adhesions and indentations caused by amniotic bands during gestation [4]. The sex ratio is equal and it is seen in all races.

Amniotic bands are usually seen during an antenatal ultrasound study or by examination of the placenta. Evidence of an embolism or an infarct may also be obtained by placental examination. There are varying hypothesis on whether amniotic bands are due to actual entanglement or due to fibrous scarring because of tissue necrosis. Amniotic adhesions may be due to secondary adhesions at the site of tissue necrosis or fibrosis [5]. Amputations of entire limbs with ring constrictions have been shown to be associated with placental evidence of amniotic strands. Although these defects can occur at any time, they usually occur during the first twelve weeks since the amnion and chorion are distinct entities during this period and the amnion is more susceptible to rupture. The formation of these bands has been associated with trauma, amniotic puncture, induced abortions and chorionic villous sampling [6]. Deformities may also be secondary to decreased

Fig. 1 : Amputation defect of right index, middle finger.

Fig. 2 : Amputation defect of digits of right foot.

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movement of a limb, as oligohydramnios is a common association. The decreased fetal activity can cause a resorptive necrosis. Vasoactive teratogens have also been implicated. Case control studies in the past have indicted a lower socioeconomic status, high parity, unwanted pregnancies and preterm births. All may be related in the sense that in lower socioeconomic status and high parity, the pregnancy may be unwanted and after an incomplete attempt at abortion, amniotic bands or emboli may form in the placenta causing infarcts. Smoking was also suggested as a causal association. Other anomalies that can be explained on the basis of amniotic band defects not seen in this case are cleft lip, syndactyly, sacral rotation and encephalocele and skull defects. In our case evidence of a constriction proximal to the distal skin tag and incomplete bands over the other limbs lead us to attribute the defect to amniotic bands. Differential diagnosis of amniotic band defects may include a syndrome of multiple benign ring shaped skin creases [7]. This is an autosomal dominant trait and these constrictions become less prominent as the child grows [8]. As for the side effects of the drugs taken by the mother in our case, metronidazole is known for its teratogenic effects in animal studies. Progestogens in pregnancy can lead to androgenic effects in female fetuses. Flouroquinolones may damage the growing cartilage. Styptics like aminocaproic acid can cause intravascular thrombosis from inhibition of plasminogen activator. The multiple drugs given to the mother could be responsible for the defect in this case.

This case report further emphasizes the importance of avoiding multiple drugs in pregnancy as these could be teratogenic. Management of amniotic band syndrome includes early detection by serial ultrasound examination during pregnancy and intrauterine interventional procedures for maintaining distal vascularity. The techniques of fetal intervention include open fetal surgery, "Fetendo" fetal surgery and fetal image-guided surgery (FIGS-IT) [9]. In open fetal surgery, the mother is anaesthetized, an incision is made in the lower abdomen to expose the uterus, the uterus is opened using a special stapling device to prevent bleeding, the surgical repair of the fetus is complete and the uterus followed by the maternal abdominal wall are closed, and the mother awakened. "Fetendo" is the name applied to fetoscopic intervention that was developed in the 1990s to avoid making an incision in the uterus and therefore, to minimize preterm labour. FIGS-IT is a term coined for fetal image-guided surgery for intervention or therapy, and describes the method of manipulating the

fetus without either an incision in the uterus or an endoscopic view inside the uterus. The manipulation is done entirely under real-time cross-sectional view provided by the sonogram. Plastic surgery will salvage the distal limb if deep grooves that encircle and limit vascular supply are treated in time. Once the distal part is gangrenous or amputated then reconstructive surgery and rehabilitation are to be carried out. Postnatally ultrasound of the brain and abdomen are important to detect structural defects due to vascular disruption. There may be hypoplasia of the lung due to oligohydramnios with respiratory insufficiency. If proper supportive care is rendered the outcome is usually good in most of the cases. Life expectancy is usually normal if visceral organs are spared. There is no risk of recurrence in future pregnancies and parents should be counselled appropriately [10].

Conflicts of interest

None identified

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