Limb Amputation Defects In Utero-A Case Report

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Abstract. Limb amputation defects in utero can be attributed to causes such as tangled strands of amnion encircling a limb or due to embolic phenomenon. Vasactive teratogens, maternal smoking during pregnancy and induced abortions are other implicated causes. We report a case in our hospital with a lower limb amputation defect and a calcaneovalgus deformity of the other foot.

Key words: Amniotic bands, limb amputation, placental emboli.

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 (Torpin 1965, Van Allen 1982, Hoyme 1981). A case of intrauterine amputation of the right lower limb is described.

Case Report:

A term small for gestational age female, weighing 2.3 kgs was born to an unbooked immunized primigravida with oligohydramnios by emergency section at our hospital. The mother and father were nonconsanguineous and were aged 24 and 26 years respectively. The pregnancy was complicated by severe abdominal pain during the late first and early second trimester for which she was given several tablets like dicyclomine, ranitidine, famotidine, metronidazole, iron and zinc preparations, hyosaminebromide, norfloxacin, aminocaproic acid, uterine relaxants like duphastan and antiemetics by a local practitioner which she had been taking regularly. There was bleeding per vaginum towards the end of the first trimester which subsided by itself. She did not undergo ultrasound examination or intrauterine procedures during pregnancy. The mother could not recollect any history of trauma. There was no exposure to radiation and the mother was non alcoholic & non smoker.

Postnatal examination of the placenta was normal. The baby had an amputation defect of the right lower limb at the proximal thigh, a laterally bent left leg with calcaneovalgus deformity of the foot. A small tag of skin separated by a band formation was present distal to the stump of the amputated right thigh. (Fig 1) There was clinodactyly of the left little finger and constriction bands on the left thigh and leg. Examination of the respiratory, cardiovascular system and abdomen were normal.

A radiograph showed a proximal part of right femur with bending of the left leg bones. (Fig. 2) The pelvis and the acetabular fossae appeared normal. There were no other skeletal defects present. Ultrasound examination of abdomen and cranium were normal, while heart revealed an ostium secundum type of atrial septal defect. The baby was referred to orthopaedics department for prosthetic device of the limb later in life and was advised to return regularly for follow up.

Discussion:

Amniotic band sequence is a set of multiple malformations due to adhesions and indentations caused by amniotic bands during gestation. The incidence is approximately 1 : 3000 newborns. The sex ratio is equal and it is seen in all races (Weidelman 1997). Amniotic bands are usually seen during an antenatal ultrasound study or by examination of the placenta. Evidence of an embolism or an infarct can also be obtained by placental examination and therefore this should be performed.

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 (Van Allen, 1981). Amputations of entire limbs with ring constrictions have been shown to be associated

Although these defects can occur at any time, they usually occur during the first twelve weeks since the amnion and chorion are distinct entities then and the amnion is more susceptible to rupture. The formation of these bands have been associated with trauma, amniotic puncture, induced abortions, & chorionic villous sampling (Van Allen 1987).

Deformities can also be secondary to decreased movement of a limb as oligohydramnios is a common association. The decreased fetal activity can cause a resorptive necrosis (Jones 1997). Aberrant bands responsible may be seen rolled up at the placental base. 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 more often the pregnancy is 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. (Aro et al, 1984). Other anomalies that can be explained on the basis of amniotic band defects, not seen in this case are cleft lip, syndactyly, sacral rotation, encephalocele and skull defects (Higginbottom 1979).

In the above 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. However since no antenatal ultrasound was done an intravascular thrombotic event cannot be ruled out. The presence of oligohydramnios in the mother supports the decreased fetal activity theory. The history of severe abdominal pain and bleeding during the first trimester are consistent with associations like threatened abortions that have been previously described.

Differential diagnosis of amniotic band defects may include a syndrome of multiple benign ring shaped skin creases. This is an autosomal dominant trait and these constrictions become less prominent as the child grows.

As for the side effects of the above drugs taken by the mother, metronidazole is known for its teratogenic effects in animal studies. Progestogens in pregnancy can lead to androgenic effects in female fetuses. Fluoroquinolones damage the growing cartilage and are therefore contraindicated in pregnancy. H2 blockers have not shown any harmful effects on the fetus however they do cross the placenta and should be given only when absolutely indicated. 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.

This report further emphasizes the importance of avoiding drugs in pregnancy since no drug is safe. The teratogenic effects of certain drugs are well known but it is better to avoid both tested and untested drugs.

Management includes early detection by serial ultrasound examination during pregnancy and intrauterine interventional procedures for maintaining distal vascularity. 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 (Bhat et al, 1991). There is practically no risk of recurrence in future pregnancies and parents should be counselled appropriately.

References :


Fig. 1
Photograph of the case showing amputation defect of the right lower limb at the proximal thigh with distal skin tag, a laterally bend left leg and a calcaneo valgus deformity of foot.

Fig. 2
Radiograph of lower limbs showing proximal part of right femur with bending of the left leg bones.