AN EXTREMELY SHORT UMBILICAL CORD WITH EXOMPHALOS

(A Case Report)

by

SUNITA PANDYA,* M.B.B.S.

and

DEVINDER SHARMA,** D.G.O., M.S.

Congenital malformations are not very uncommon. There are the minor ones which pass unrecognised. It is rare for the major deformities to cause dystocia, because they are often associated with abortion or premature labour. With the early diagnosis of a gross deformity treatment is simple, while with delayed diagnosis there may be a grave risk to the mother or severe injury to the lower genital tract due to forcible extraction.

The incidence of congenital deformities is difficult to estimate. Some deformities are so severe as to cause death in utero, others are compatible with survival for a few hours or days, others are incompatible with life unless special surgical treatment is carried out in the neonatal period and yet others are so trivial as to present no threat to survival.

Following is a case of a congenital malformation in a foetus associated with severe degree of placenta praevia and an extremely short umbilical cord.

Case Report

M.D., 28 years old multigravida was admitted on 15-3-71 at 8.45 P.M. with the complaint of amenorrhoea for seven months, bleeding per vagina for three days and pain in the abdomen for one day. She had previous four full term normal deliveries (Two alive, two died). The last delivery was two years back. Her menstrual history was normal. There was no history of any significance in any of the previous trimesters.

On examination, the general condition of the patient was poor. She was of thin built with pallor of moderate degree, pulse 140/min. regular, B.P. 100/70 mm of Hg. and temperature 101°F with dehydration. On systemic examination nothing abnormal detected.

On abdominal examination, the fundal height was 28 weeks, LOA vertex presenting, mild uterine contractions present. No uterine tenderness. Foetal heart sounds were present 180/min. regular. Blood was arranged and an intravenous drip was started. Patient was given 1/6th gr. morphine sulphate intramuscularly immediately after admission to combat shock and allay her anxiety. As the patient presented with antepartum haemorrhage the operation theatre was prepared for caesarean section. On examination in the theatre the uterus was found to be actively contracting. Since the patient was in active labour, and there was slight bleeding, a vaginal examination was done. The cervix was more than three fifth dilated, fully taken up, membranes were absent and a hand was found hanging through the dilated os. The edge of placenta was found anterolaterally. The head was

*Clinical Registrar in Obstetrics and Gynaecology, Institute of Medical Sciences, Banaras Hindu University, Varanasi-5.

**Reader in Obstetrics and Gynaecology, Institute of Medical Sciences, Banaras Hindu University, Varanasi-5.

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high up towards the right side in the brim. Since there was no active bleeding and baby also very premature to survive, it was decided to avoid an abdominal delivery.

Under deep general anaesthesia internal podalic version was done without any untoward difficulty and gently breech extraction was performed. During breech extraction some boggy mass was felt on the foetal abdomen, which was later on discovered as a major degree of exomphalos covered with membranes only. Placenta delivered immediately after breech extraction. Injection methergin, one ampoule, intravenous was given stat. There was a mild postpartum haemorrhage. Patient had an uneventful puerperium. There was no sepsis as prophylactic was given. Patient was discharged on fourth day with antianaemic treatment, as her haemoglobin was 6 gms% on discharge.

On examination of the foetus, major exomphalos covered with membranes with a very short cord was found. Both the lower limbs were extended at the hip joint as well as at the knee joint and lying posteriorly. External genitalia were formed, it was a male foetus. It was a fresh stilbirth, weighing two pounds two ounces.

On examination of the placenta, it was comparatively bigger in size, very friable, not healthy looking, cotyledons were oedematous; cord was inserted eccentrically on the foetal surface of the placenta. Cord length was one inch (2.5 cm.) approximately, weight of placenta was two pounds. Thus, foetal placental ratio was 1:1. The foetus and the placenta were sent for histopathology to know further details and to exclude other deformities.

Histopathological Report

Six months old foetus with a membrane covered mass projecting from the umbilicus (12 cm.). On cutting the abdominal wall it was deficient and there was protrusion of liver, loops of intestines and kidney. Small cord (2.5 cm.) attached to the upper portion of the mass. Placenta was discoid, spongy, grey in colour. Liver showed extramedullary haematopoesis. Placenta and other organs did not show any remarkable pathological change.

Comments

Malpas (1937) found 294 malformations in 13964 total births in Liverpool Maternity Hospital, an incidence of 2.1 per cent. Logan (1951) reported an incidence of 2.5 percent. McKeown (1960) published a record of a survey of 56760 births in Birmingham. He collected data from doctors and midwives of children two weeks after birth and followed 80 per cent of these upto five years. The incidence of deformities in babies within two weeks of birth was 17.3 per 1,000 total births. Total number of malformations estimated after five years was 23.8 per 1,000. Since more malformations come to light as the child gets older, the observed incidence upto five years was higher than at birth. These are, therefore, defects which are compatible with life and are easily missed at birth. Some deformities may pass unrecognised unless a postmortum examination is performed, when a baby dies of pneumonia or any other disease.

Exomphalos, hernia of abdominal contents, when it occurs in a minor form is compatible with life if an early surgical repair is done. When it is of a major degree i.e. an infant is born with a large hernia covered with amnion, practically all the intestines and usually a part of the liver lie outside the shrunken abdominal cavity; the umbilical cord is asymmetrically attached to the apex of the amnion with a gross deficiency of the abdominal integuments. Exomphalos of such major degree is incompatible with life. The baby usually dies of other associated abnormalities or because of local infection. It is reported that 20% of cases of exomphalos have major cardiac lesions and 5% of cases are associated with ectopia vesicae.

The shortest cord length reported by

Eastman was 0.5 cm. and the longest cord reported was 198 cms. It has been shown that when the placenta is situated in the fundal area, the minimal length of the cord which will allow normal vaginal delivery of the child without undue traction on the cord is about 35 cm. and when the placenta is in the lower uterine segment the minimum length of the cord required is 20 cms. (Eastman & Hellman).

Shortness of the cord, whether absolute or relative, may cause malpresentation and position of the foetus, premature separation of the placenta, premature rupture of membranes, prolonged labour, rupture of cord, intrauterine foetal death, exomphalos and rarely inversion of the uterus (as in Dyhenfurth's case, 1911). Absolute shortness of the cord below 8″ (20 cm.) is rare except in association with foetal malformation, notably exomphalos (Brews and Bender). Similar case to this has been reported by Rajpal and Mallik (1966), the length of the cord in their case was 2″ (5 cms.). In our case the length of the cord was 1″ (2.5 cms.).

It is difficult to comment on the association of severe degrees of placenta praevia in such type of cases. Since the placental site is determined prior to the development of the cord and the rest of the foetus it is obvious that the shortness of the cord is not as a result of placenta praevia.

It is known that herniation of abdominal viscera is a normal condition in human embryos between the 10 mm. (end of fifth week) and the 40 mm (third months). The rapidly growing liver and the developing mesonephros encroach on the available space in the coelom so much that the U-loop of intestines is extruded into the portion of the extra embryonic coelom which becomes included in the umbilical cord. The part of the extraembryonic coelom included in the umbilical cord acts as a sac for the normal umbilical hernia which characterise the embryo between sixth and the tenth week. After the disappearance of this hernia the remaining of the extraembryonic coelom normally becomes obliterated. Under abnormal conditions, it may persist as exomphalos at birth.

Summary

An interesting case of an extremely short cord associated with exomphalos and major degree of placenta praevia is presented with the review of literature.

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