SEVERE FORM OF ADAM COMPLEX - INTRANATAL MALFORMATION

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ABSTRACT

ADAM complex (Amniotic Deformities / Adhesions / Mutilations) is a rare poly-malformation syndrome with an incidence of 0.33 for every 1000 births in the literature with thoraco- and/or abdomino-schisis associated with an eventration of the internal organs and variable variations in the extremities. The present foetus diagnosed with ADAM complex showed placenta forming an operculum at the cranial pole, severe craniofacial defects, absence of right upper limb and malrotation of right lower limbs, complete malrotation of the circulatory, respiratory and gastro-intestinal systems .The novel thing in this case is the presence of malrotation which has not been reported before. The cerebral hemisphere was partially present. A rudiment of brain stem without any differentiation was present. The cerebellum and spinal segments were absent. Postmortem foetogram revealed undifferentiated components of axial skeleton. The mechanism responsible for the Adam complex is the presence of Amniotic Band during early embryonic period.

Key words: ADAM complex, thoraco/ abdominoschisis, malrotation and Amniotic Band

INTRODUCTION

ADAM complex (Amniotic Deformities / Adhesions / Mutilations) complex[1] is a severe form of Amniotic band sequence. Amniotic bands were first mentioned by Hippocrates (B.C), described by Chaussier (1800) and emphasized by Higginbottom (1979) that the severity of the anatomical variations are directed by the developmental age at which the disruption of the amniotic membrane occurs[2]. The adherence of amniotic bands on the embryonic disc yields polymalformations which include circulatory , respiratory, central nervous system ,gastro-intestinal and ventral abdominal wall. Rosso(1993) differentiated two phenotypes placento-cephalic and placento-abdominal by the attachment of the placenta to the cephalic end and abdominal walls respectively[3]. Allen et al. in 1987 while describing limb body wall complex propounded a three point diagnostic criteria : 1. Thoracic and/or abdominal coelosomia. 2. Exencephaly or encephalocele with a facial cleft. 3. Anomalies of the extremities to ascertain the severity[4]. The present case expressed all the three criteria to be considered as the severe form of the amniotic band syndrome.

CASE REPORT

A 29 year female spouse with non-consanguineous marriage, G2P1L1A0 with a healthy first child, and no family history of congenital variations, underwent medical termination of pregnancy as the antenatal obstetric ultrasound scan revealed gross anatomical variations i.e., a lump of tissue with undifferentiated trunk, neck and head with only 3 limbs and bad prognosis. An 18week stillborn (according to last menstrual period) female foetus was delivered with the following features.

Post mortem foetogram (fig. 1) showed undifferentiated skeletal elements in the axial region. Right upper limb was absent and the left upper limb bones were located close to the lower limb bones. The lower limbs showed normal position of femur but right tibia was curved and twisted posteriorly so that the entire foot was positioned posterior. Fibula was absent in both the lower limbs. Cranio-facial defects: Cranio-placental attachment (fig.2) and the amniotic membrane extending onto the thoraco-abdominal region, plastering the umbilical cord to the amniotic membrane was observed (fig. 3). Presence of cyclopean eye, and absence of nose, oral fissure and neck were observed (fig. 4).

After detaching the placenta, cranial vault was found to be absent with persistence of brain tissue. Anterior wall of trunk was absent in the supra umbilical part and was covered by amniotic membrane and the infra-umbilical part of anterior abdominal wall was normal. Coils of intestine were present in the cephalic pole of the trunk and liver was present in the caudal pole of the trunk. Cardio-vascular, respiratory and gastro-intestinal systems were completely malrotated, this being an unique feature of this case having not been reported

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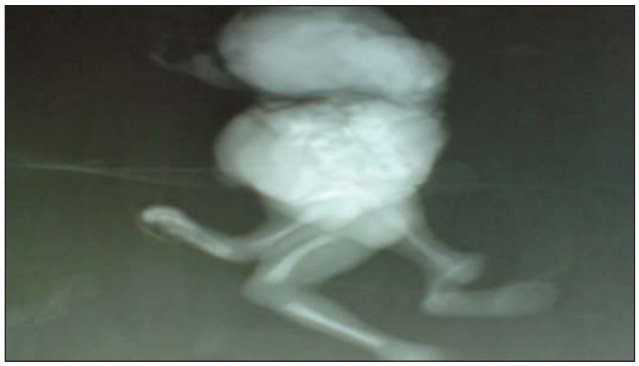

Fig.1- Showing foetogram of foetus with ADAM complex.

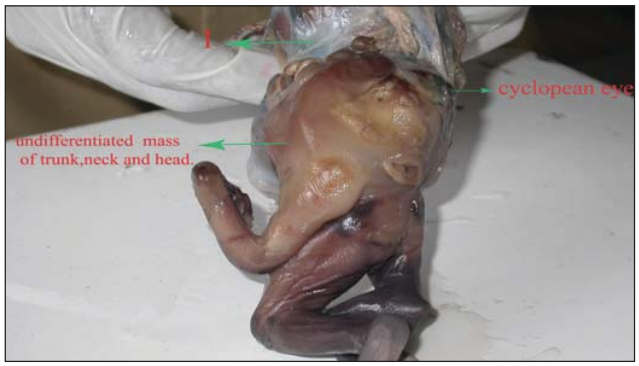

Fig.2- Showing Cranio-placental attachment, Cyclopean eye and undifferentiated mass of trunk, neck and head.

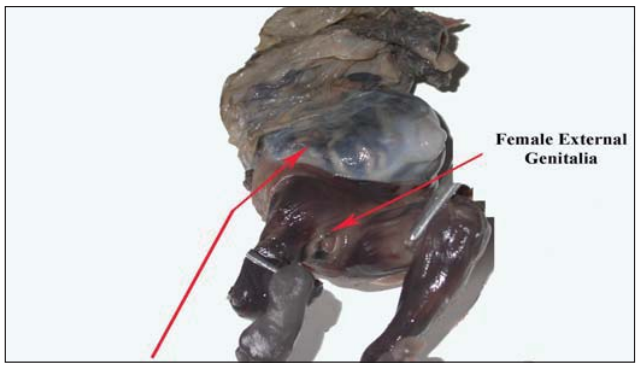

Fig.3- Showing amniotic membrane extending onto thoraco-abdominal region.

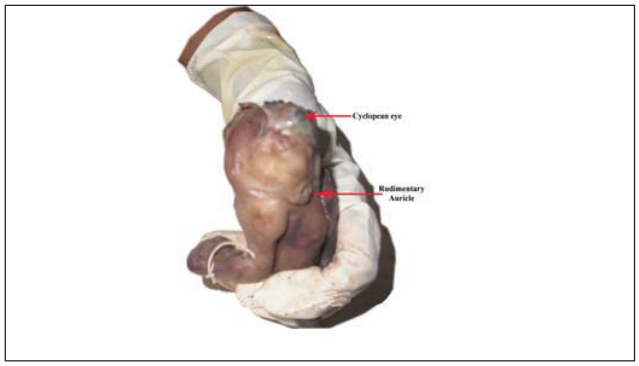

Fig. 4 : Showing cyclopean eye and rudimentary auricle, absence of nose, oral fissure and neck.

before. After dissection the liver, heart and lungs were exposed which occupied entirely the left side of the body in a linear position. As such, in this present case the lungs have been termed as upper and lower lungs with respect to the position of the heart. The cardiac and pyloric ends of the stomach were rotated upside down. Kidney was unilaterally present on the right side and absent on left side with presence of bilaterally placed suprarenal glands. Uterus was cord like with a streak ovary on the right side. Karyotype of the foetus was 46 xx.

DISCUSSION

Many authors, in their studies of ADAM complex reported cephalo-placental attachment with craniofacial variations, thoraco and abdominoschisis and limb variations. However malrotation of systems associated with ADAM complex has not been reported. Rare foetal polymalformations of uncertain etiology as in the present case, have been regarded as sporadic in nature with a low recurrence risk. Cases of constrictive amniotic bands and cranioplacental attachment, have been reported[5].

H. Saadi, K. Sfakianoudis & D. Thomas (2007) in their study of Limb body wall complex associated with placenta previa accreta reported an anterior body wall defect with evisceration of the heart, stomach and bowel[6]. Skeletal anomalies included hypoplasia of the left forearm with only one bone fragment being present, hypoplasia of the hand with absence of fingers and apparent kyphoscoliosis. The present case manifested malrotation of multiple systems as additional complication. Amongst the three theories of empirical understanding i.e., Exogenic theory, Vascular theory described by Van allen (1987)[4] and Streeter's theory-developed by Hartwig (1989)[7], Exogenic theory propounded by Torpin et al. (1965)[8] based on early rupture of amnion leading to amniotic bands that interrupt embryogenesis resulting in a wide range of variations of number of systems influencing various genes simultaneously is more self explicable. Moerman P[9] conducted foetopathologic evaluation of amniotic bands and described three types of lesions (i.e.) constrictive tissue bands, amniotic adhesions and limb body wall complex with increasing complexity. Malrotation of the viscera of thoracic cavity, abdominal cavity and cranial cavity a unique feature of the present case adds much gravity to the fatal nature of the deformity, implying medical termination pregnancy as an ascertained eventuality at the earliest diagnosis of the condition. As ADAM complex is not genetically predetermined,

recurrence in the following pregnancy would also be sporadic and not ascertained.

CONCLUSION

Further refinement of the diagnostic technique; evolving a protocol considering the finer details of the sonological findings of this condition and noninvasive biochemical investigations as the prenatal diagnostic tools, is the need of the hour to be able to detect the more obscure amniotic bands as early as possible either to intervene or be precisely decisive of the prognosis and offer the perfect advice to the pregnant women.

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