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Case Study

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PLACENTO-CRANIAL ADHESION SYNDROME: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Direct attachment of placenta to the fetal skull is a rare condition generally included under amniotic band syndrome. Based on shared features of the four cases in the past, we had proposed a new syndrome comprising of placental attachment, skull defect, anencephaly, facial dysmorphism, short umbilical cord, low weight for gestation and oligohyramnios. In addition nearly all the cases were fatal. We are presenting autopsy findings of the fifth case which shares all the features except anencephaly instead of which scizencephaly is observed. There is also bilateral mesomelia, micropthalmia and abdominoschisis. We have justified distinguishing this condition from amniotic band syndrome and limb body wall complex.

KEYWORDS: Placento-cranial; amniotic band syndrome; limb body wall complex; syndrome.

INTRODUCTION

Cases of placento-cranial adhesions (PCA) where in placenta is directly attached to the fetal skull are rare. There is no consensus regarding appropriate classification of such cases and often in literature these have been included under either amniotic band syndrome (ABS)/ ADAM complex (amniotic deformity, adhesion and mutilation) or limb body wall complex (LBWC)/ body stalk anomaly (BSA).[1] Till date the largest series of PCA has been reported by us wherein we had carried out detailed autopsies of four cases all of which had died in utero. Based on features present in all four cases i.e. placento-cranial adhesion, skull defect, anencephaly, short umbilical cord, low weight for gestation, facial dysmorphism, limb/digit amputation and oligohydramnios we had proposed a new syndromic association.[2] We are presenting a fifth such case with all the features except anencephaly instead of which schizencephaly is noted. In addition there is abdominoschisis, bilateral hypoplastic radius and ulna and micropthalmia. In light of the present case and after perusal of literature we are modifying the features of PCA syndrome and also offering valid reasons for differentiating PCA from ABS/ADAM and LBWC/BSA.

Case

A 31 years old primigravida had registered for antenatal care. Around 19 weeks of gestation, on ultrasound examination abdominoschisis was detected. At 21 weeks of gestation labor was induced after a diagnosis of intra uterine death was made. There was no history of consanguinity or abnormal drug intake by mother. Ultrasound had failed to detect the anomalies however oligohydramnios was noted. A male fetus weighing 220 grams with crown rump length of 15 cm was delivered and sent for complete autopsy. Placenta was directly attached to the skull and umbilical cord length was 17 cms. The underlying skull defect involved partial absence of parietal and occipital bones. The brain had open schizencephaly with occipital cleft accompanied by partial absence of sulci and gyri. The cerebellum appeared to be normal. There was bilateral micropthalmia and nose was bifid. Abdominoschisis was noted. Liver along with most of the intestines were out of abdominal cavity. Bilateral radii and ulna were hypoplastic, qualifying for upper limb mesomelia and there was oligodactyly with one of the fingers missing on both hands. The thoracic cavity contents however in the normal anatomical locations with bilateral pulmonary hypoplasia. Lower limbs showed bilateral talipes equinus varus.

Fig. 01.

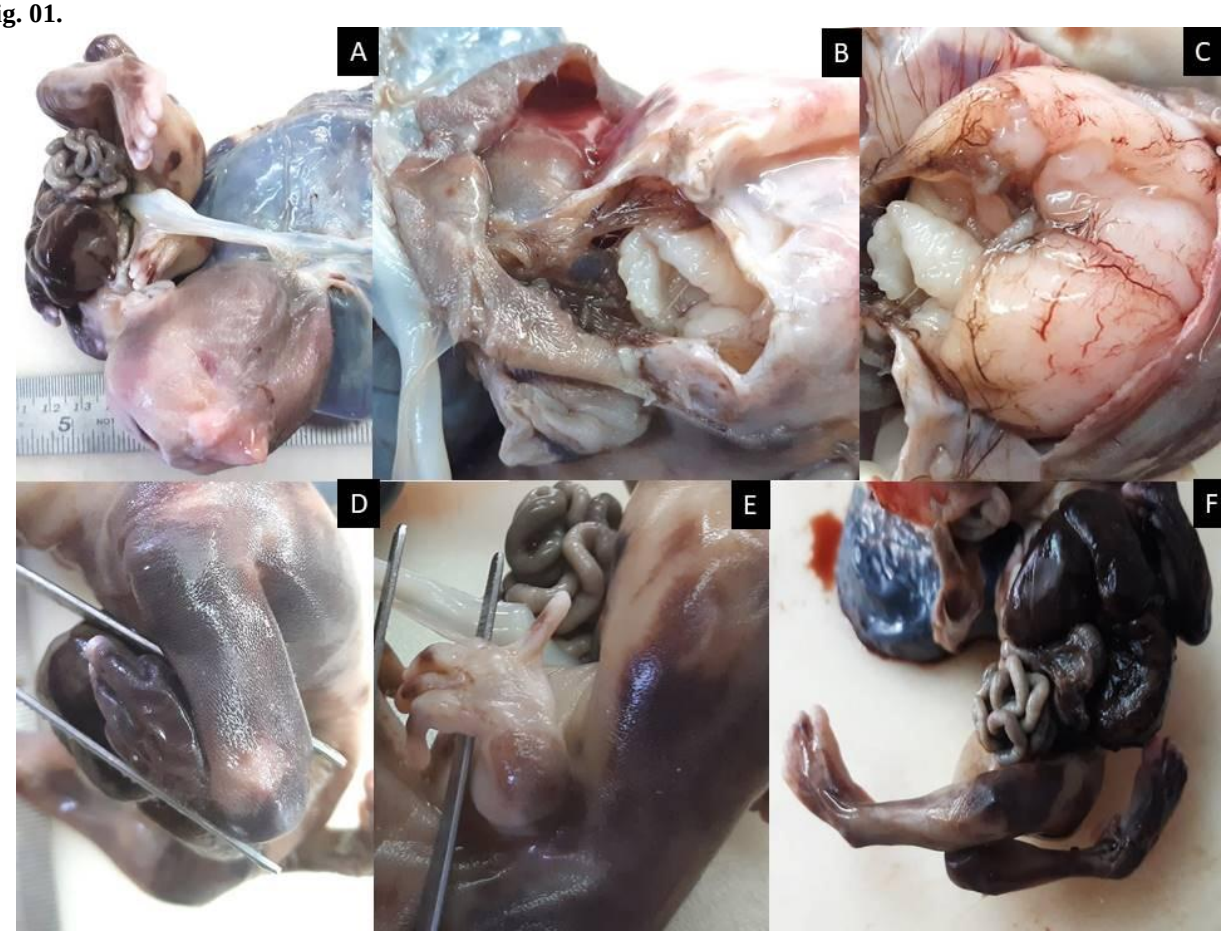

Fig. 01: A, Placental attachment to head along with bifid nose and micropthalmia; B, Parts of occipital and parietal bones missing; C, Underlying open schizencephaly; D, Mesomelia ; E, Oligodactyly; F; Bilateral CTEV and abdominoschisis. Liver and intestinal loops visible.

Table 01: Fetal features compared with expected features at the gestational age.

Gestational age Weeks	Crown rump length (cm)	Expected age corresponding to CR length (days)(3)	Umbilical cord Length (cm)	Expected mean umbilical cord length(4)	Weight	Expected weight(3)
21	15	123	17	32	220	400

DISCUSSION

Classification of cases such as presented here is fraught with problems since apparently there is no guiding consensus in literature. Various authors have included placento-cranial adhesions (PCA) under the rubric of amniotic band sequence/syndrome (ABS). **(5)** ABS is characterized by amniotic bands resulting in disruptions including craniofacial malformations with or without amputations, body wall defects and visceral anomalies.[6] LBWC can be considered as a part of ABS owing to overlapping features. Rationale behind separation of LBWC from ABS rests on the essential criteria of body wall defect and extrusion of organs in the former and therefore serious implications whereas the spectrum of ABS might range from mild features such as cleft lip to major disruptions like body wall defects.[1]

It can be argued that PCA cases similar to the one presented here can be classified as limb body wall complex since the latter includes exencephaly/encephalocele with or without craniofacial defect, body wall defect in the form of thoraco and/or abdominoschisis and limb defects.[1] Russo et al have classified LBWC into two phenotypes; placenta-cranial and placento-abdominal adhesion with craniofacial abnormalities in former and none in the latter.[7] Our case therefore would straddle the two phenotypes according to Russo since there is both abdominal wall defect as well as placenta-cranial adhesion.

Classification or categorization of any disease entity is largely based either on etiopathogenesis including genetic basis or phenotype which dictate the management and therefore the prognosis. From etiopathogenesis point of view there is no discernible evidence to pigeonhole ABS, LBWC or PCA into different categories provided placento-fetal adhesion is a consequence of same sequence of events as ABS which

being damage to the amnion early in the pregnancy resulting in amniotic bands with or without placental attachment as initially proposed by Torpin et al[8] However different phenotypes with respective prevalence and prognosis do offer compelling reasons for further classification. Perusal of literature shows that ABS is most common, followed by LBWC and least frequent is PCA.

ABS or ADAM complex (amniotic deformity, adhesion and mutilation) denotes an overarching wide spectrum of phenotypes with prevalence of 1 in approximately 11,000 live births.[9] The prevalence of LBWC on the other hand appears to be 1 in 7500 at 10-14 weeks of gestation.[10] It must be noted that prevalence of ABS is based on live births whereas LBWC is irrespective of viability of pregnancy. Some authors have quoted the prevalence of LBWC ranging from 0.21 to 0.31 per 10,000 viable deliveries.**[11]** There is no reliable data on the prevalence of PCA however in our centre it is 5 per 10,000 pregnancies.

Regardless of prevalence there is consensus apropos survival. Most cases of LBWC are fatal with a few exceptions where it has been possible to save the neonates.[12] On the other hand there is no single case report detailing long term survival of PCA although there are reports of live births.[13] In our centre all four previous cases and the present one had died in utero between 16 to 21 weeks of gestation.[2]

Therefore the trifecta of placenta-cranial adhesion, skull defect and brain malformation resulting in fetal or neonatal death itself should suffice for a separate class differentiating it from LBWC. A shown in table 1, all such cases show at least three of the above mentioned features namely placentalo-cranial attachment, underlying skull defect and brain anamoly along with facial dysmorphism. It is interesting to note that short umbilical cord is a finding associated with all the cases of PCA in our experience as well as LBWC in literature.[14] Fig **02** Venn diagram put in perspective, the relationships between different entities.

Fig. 02.

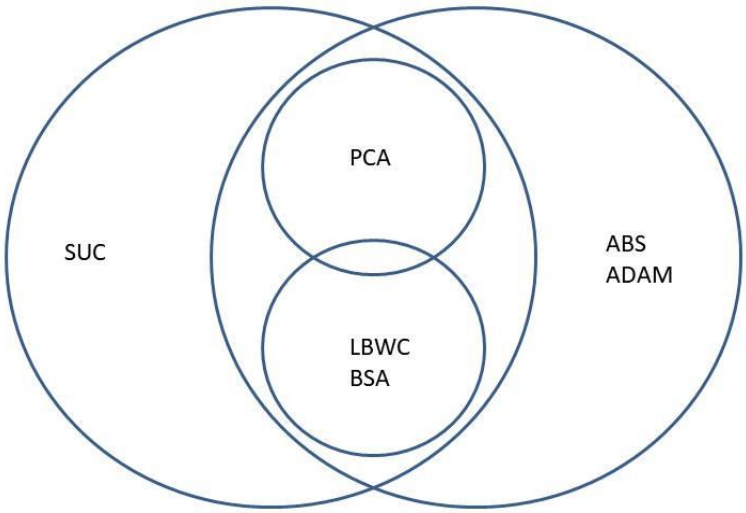

Fig. 02: Venn diagram showing relationship between PCA (Placento-cranial adhesion), LBWC (Limb body wall complex)/BSA (body stalk anomaly), ABS (amniotic band syndrome)/ADAM (amniotic deformity, adhesion and mutilation) and SUC (short umbilical cord).

Table 02: Cases of PCA with features and outcomes with references Y: Present; N: Not present; NA: not available.

Reference	Skull defect	Brain	Facial Dysmorphism	LBW	Short UC	Oligo Hydramnios	Amputation	Nasal cleft	Abdominal wall defect	Cleft lip/palate	Outcome
[5]	Y	Schizencephaly	Y	Y	NA	NA	Y Digits	N	N	Y	36 wks Operated Died
[15]	Y	No major changes	Y	N	NA	NA	Syndactyly	N	N	N	31 wks Operated FU
[16]	Y	+	Y	NA	NA	NA	N	N	N	Y	20 wks
[17]	Y	+	Y	Y	NA	CTEV	Y	N	N	Y	36 wks Hypoplasia radius/ulna. Anopthalmos.
[17]	Y	+	Y	Y	NA	N	N	N	N	Y	Preterm
[18]	Y	+	Y	Y	Y	CTEV	N	Saddle nose	N	Y	28 wks
[2]	Y	Anencephaly	Y	Y	Y	CTEV	Y Digits	Cleft	N	Y	16
[2]	Y	Anencephaly	Y	Y	Y	CTEV	Y Digits	Cleft	N	N	20
[2]	Y	Anencephaly	Y	Y	Y	CTEV	Y Digits	Cleft	N	N	20
[2]	Y	Anencephaly	Y	Y	Y	CTEV	Y Digits	Cleft	Y	N	21
Present case	Y	Schizencephal	Y	Y	Y	CTEV	Y Digits	Bifid	Y	N	21

PCA can be partly described by sequence of events following premature rupture of amnion and consequent amniotic band formations including placental attachment to the skull.[19] Shear mechanical forces as a consequence of placental attachment might explain facial dysmorphic features whereas CTEV can result from reduced fetal movements owing to oligohydramnios.[20] It is difficult to explain with same degree of simplicity and clarity the features seen in PCA. Short umbilical cord can be a result of reduced fetal movements.[21] Oligohydramnios and brain anomalies can also result in decreased fetal movements.[22] It is plausible that placental attachment to the skull with pre-existing bony defect is facilitated by sluggish fetal movements. The underlying brain defect can be explained by possible erosive action of MMP2 and 9 secreted by placenta on the brain.[2]

We maintain that direct attachment of placenta to any fetal part is almost always accompanied by an anatomical defect at the point of attachment. It is difficult to ascertain whether anatomical defect results in placental attachment or vice versa.[2]

Features in the present case such as schizencephaly, upper limb mesomelia and micropthalmia have not been reported in the literature in association with PCA and as such are difficult to explain in context of ABS or feto-placental adhesion.

Given the fact that all cases of PCA are accompanied by significant brain anomalies even on gross examination, the surgical correction of live births are likely to be complicated by permanent compromised centre nervous system functionality in contrast to placenta-abdominal adhesions or LBCW wherein brain and other craniofacial features are normal.

There is no gain saying that some cases, like the present one are going to have features of both placenta-cranial adhesion and LBWC, nevertheless it would be logical to classify them under PCA since the brain, the principal prognosticator of survival with quality of life in such cases is abnormal. In the light of our experience with cases of PCA and relevant published literature thus far, it would not be injudicious to terminate such pregnancies. The purpose of reporting present case is to add on to literature and also demonstrate importance of fetal autopsies.

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Disclosures: None.

CONFLICT OF INTEREST: None.

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