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CASE REPORT

DIPROSOPUS TETROPHTHALMUS INCLUDING RADIOLOGICAL AND POST-PARTUM IMAGES-A CASE REPORT

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ABSTRACT

Conjoint twinning is very uncommon with an incidence of approximately 1:50,000 to 1:100,000. Diprosopus tetrophthalmus is the rarest variant of conjoined twinning which results from incomplete monozygotic, monochorionic and monoamniotic twin division associated with multiple congenital malformations.A 25 years old female patient at 26 weeks of gestation having single gravida (primi) and no significant family history was referred to our radiology department for anomalies scan. Transabdominal obstetric ultrasound showed the fetus with one head, two faces, two legs, two hands, one thorax, one abdomen and a single spinal cord with deformed spine.The aim of this case report was to present and discuss a case of Diprosopustetrophthalmus (craniofacial duplication) a rare variant of conjoined twins resulted from incomplete monozygotic, monochorionic and monoamniotic twin division which help in prenatal counseling to the patient and its management.

Keywords: *Conjoint Twin, Diprosopus Tetrophthalmus, Monocephalic*

INTRODUCTION

Diprosopus tetrophthalmus is the rarest variant of conjoined twinning which results from incomplete monozygotic, monochorionic and monoamniotic twin division associated with multiple congenital malformations[1,2]. Diprosopus tetrophthalmus is a rarest type of conjoined twins with incidence reported from craniofacial duplication or Diprosopus 180000/15000000 [6].

Although its etiology is not known well, they have a wide spectrum from double nasal structure to double face, four eyes (tetrophthalmos) and single head (diprosopus monocephalus)[9]. We report a case of Diprosopus tetrophthalmus which help in prenatal diagnosis and subsequently help in prenatal counselling to the patient party and its management.

CASE SUMMARY

A 25 years primi at 26 weeks of gestation presented to radiology department, National Medical College, Birgunj for anomalies scan having no significant complaints, no positive family history, and no history of smoking or drinking or drug abuse. 2D, 3D and 4D ultrasound was performed. Obstetric ultrasound report showed single live fetus around 26 weeks of gestation with cephalic presentation, polyhydramnios and multiple congenital anomalies along with one head, two faces, two legs, two hands, one thorax, one abdomen, a single spinal cord, deformed spine.

Fetal head scan showed single enlarged head with severe hydrocephalus and compressed surrounding brain parenchyma. Falx was central with partial fusion of posterior fossa structure seen. Calvaria also appeared to be normal.

Facial scan showed two facial structures with four eyes; two in the mid line closely situated and two laterally placed with four eye balls. Two mouth openings with two nose both had cleft lips and cleft palates. One nose consisted of nasal polyp. However, nasal bones appeared normal.

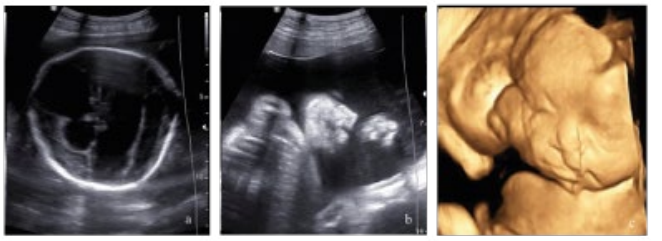

Figure 1:a) Grey-scale ultrasonographic image showing hydrocephalus b) cleft lip c) 3D ultrasonographic image showing cleft lip and diprosopus

On body scan there was a single body with normal bilateral upper and lower limbs. There was normal bony anatomy of bilateral upper and lower limb including pelvis. Umbilical cord appeared to be encircled around the right thigh.

Fetal spine scan in sagittal, coronal and axial view showed slightly deformed and misalignment of the vertebral bones and deformed cranio-cervical junction. There was duplication of cervical, thoracic and lumber vertebra with splaying of vertebral pedicles at lumbar region and mass outpouching through the splayed pedicles consisting of CSF and cauda equina covered by skin and meninges consistent with lumbar meningomyelocele.

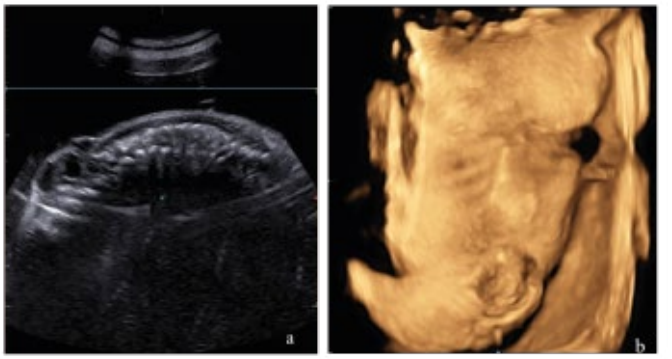

Figure2: a) 2D ultrasonographic image showing lumbar meningomyelocele b) 3D ultrasonographic image showing lumbar meningomyelocele

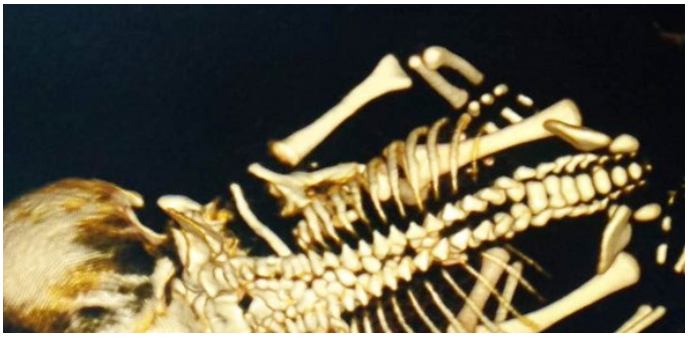

Figure 3: CT 3D Reconstruction image showing large skull with spinal duplication.

There was one thorax with normal situs consisting of normal ribs with two normal lung tissues, one trachea and esophagus. There was single heart with normal heart rate consisting of 4 chambers, normal aorta and venacava. However, small VSD was noted.

Fetal abdominal scan showed one abdomen with normal situs of abdominal organs consisting of normal bowel loops, stomach, bilateral kidneys and urinary bladder. No anorectal malformation detected.

On post-partumthe baby was found to have two faces with four eyes, two laterally placed and two in the midline. There were two mouths and two noses with cleft lip and palate. There was one thorax, one abdomen and two upper and lower limbs with normal phalanges. The baby was male with normal genitalia.

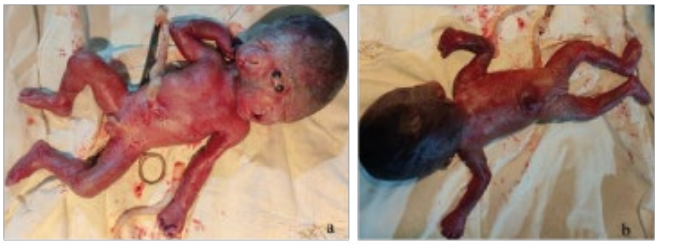

Figure 4: Post-partum photograph a) Diprosopus Tetrophthalmos b) Lumbar Meningomyelocele

DISCUSSION

Diprosopus tetrophthalmus is the rarest variant of conjoined twinning which results from incomplete monozygotic, monochorionic and monoamniotic twin division associated with multiple congenital malformations[1,2]. Less than 1% of conjoined twins have diprosopus, which can range from minimal facial structure duplication to complete dicephalus[1,2]. It is characterized by a wide spectrum of abnormalities that range from simple nasal duplication to two complete faces with a single body.

Tetrophthalmus refer to four eyes, can range from four fully separate globes and bony orbits, to two separate globes laterally with partial or full midline orbital fusion [2] as in our case. Duplication of eyes is always associated with duplication of nose as in our case but duplication of nose may be isolated[5]. We could not conduct a chromosomal analysis in our case since the parent did not allow.

Several other abnormalities associated with Diprosopus have been described in the CNS, CVS, GI and respiratory systems, and most neonates are stillborn[5].The more extensive duplications are associated with more severe central nervous system anomalies. Brain abnormalities include complete or partial duplication of the cerebral

hemispheres with varying degrees of fusion of the posterior fossa structures[6,7]but in our case, there waslive fetus of 26 weeks of gestation with bilateral hydrocephalus, partial fusion of posterior fossa structure and there was no duplication of cerebral hemisphere.

Imaging of diprosopus includes 2D and 3D sonography performed prenatally to show the features of facial duplication [2, 6, 8] as in our case there is two separate nose, face and mouths along with cleft lip and palate in both sides.

Polyhydramnios is common finding probably as a result of difficulty in amniotic fluid swallowing as in our case there was polyhydramnios.

Multiple spinal abnormalities, including thoracic and cervical duplications, have been observed[15] as in our case there was duplication of cervical, thoracic and lumber spine with lumber meningomyelocele.

Other organ defects include diaphragmatic hernia,VSD,overriding of aorta, hypoplastic aorta and dextrocardia, dysplastic cystic kidneys, hypoplasia of urinary bladder, cleft lip, palate and imperforate anus[9] but in our case, only VSD was noted. In our case, bilateral kidneys, diaphragm and bowel loops appeared normal with normal abdominal and thoracic wall.

CONCLUSION

Due to advance imaging modalities in recent days, early detection of congenital anomalies with conjoint twins such as Diprosopus tetrophthalmus is essential in term of social, economic and ethical aspects and it will enable patients to decides at early weeks.

CONSENT

An informed and written consent was obtained from the mother for publication of clinical details and images.

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