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 Diprosopus tetrophthalmus (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. Diprosopus tetrophthalmus is a rarest type of conjoined twins with incidence reported from craniofacial duplication or Diprosopus 180000/15000000.

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). 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 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 Diprosopus tetrophthalmus (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
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 lumbar 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.

On post-partum the 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.

**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. Less than 1% of conjoined twins have diprosopus, which can range from minimal facial structure duplication to complete dicephalus. 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 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. 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. 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 but in our case, there was live 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 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 as in our case there was duplication of cervical, thoracic and lumbar spine with lumbar 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 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 decide at early weeks.

CONSENT

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

REFERENCES


