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Case Report

Prenatal diagnosis of the rarest conjoint twin "diprosopus tetrophthalmus" during anomaly scan: A case report ☆

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ABSTRACT

Twin pregnancy is considered high-risk pregnancy because of its various effects on maternal and fetal physiology. Twin pregnancy can be dizygotic or monozygotic, the latter being less common. Depending upon the period of separation of an embryo, monozygotic twins can share amniotic cavity and placenta with the monochorionic monoamniotic form being the least common type. Diprosopus tetrophthalmus is the rarest form of monozygotic monochorionic and monoamniotic conjoint twin, where there are 2 faces on 1 head with various degrees of duplication of facial and cranial structure. The exact etiology of diprosopus is still unknown however; there are many local environmental and oxidative theories for this anomaly. The incidence of diprosopus is 1 in 180,000 to 15 million births. Here we present a case of 20-year-old female who came for routine anomaly scan of her pregnancy in our hospital which showed the presence of diprosopus tetrophthalmus. Couples agreed to terminate pregnancy after proper counseling from treating physician. Ultrasonography images of anomaly scans as well as post-abortion images of the fetus are discussed in this case report.

Introduction

Twin pregnancy can be dizygotic where 2 ova are fertilized by 2 sperm or monozygotic where 1 ovum is fertilized by 1 sperm. Depending upon period of separation of an embryo during pregnancy, the monozygotic twin can share placenta and amniotic cavity with resultant dichorionic diamniotic, monochorionic diamniotic and monochorionic monoamniotic types of twin pregnancy. Diprosopus is the rarest type of conjoint twin belonging to the monochorionic monoamniotic group with a reported incidence of 1 in 180,000 to 15

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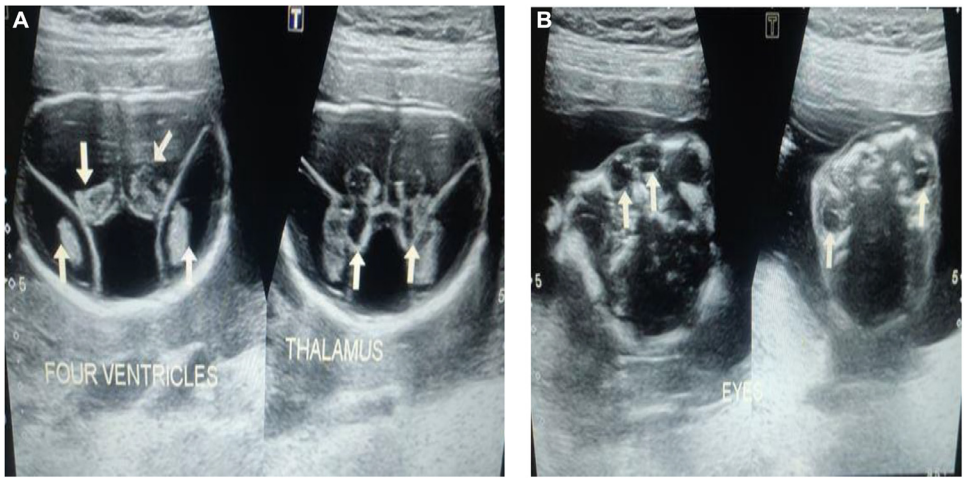

Figs. 1 – A and B: Axial B-mode ultrasound image of skull showing dilated and duplicated bilateral lateral ventricles with double thalamus (A) and thinned out brain parenchyma (white arrows). Skull vault is absent in anterosuperior aspect however, it is present posteriorly. Second image (B) showing presence of 4 eyes with 2 in midline and 2 placed laterally suggestive of tetrophthalmus (marked by white arrows).

million births [1]. There is a characteristic presence of 2 faces on a single head and trunk with variable degrees of duplication of facial and cranial structure [1]. It is commonly associated with anencephaly, spinal bifida, and various other cardiac and skeletal malformations. Anomaly scan in the expert hand plays a vital role in prenatal diagnosis of this entity. Here we present a case of diprosopus tetrophthalmus with spinal bifida and bilateral club foot diagnosed at 17 weeks of period of gestation.

Case presentation

A 20 year female G1P0 at 18 weeks of period of gestation came for her routine follow-up examination of her pregnancy. Her medical history was unremarkable with no history of intake of any drug other than iron and calcium for her pregnancy. She took vitamin folic acid during the preconceptional period and in the first trimester. She was non-alcoholic and non-smoker. On clinical examination, her vital was stable with unremarkable clinical examination. Her routine lab parameters including complete blood count and serology were within normal limits. Her dating scan was done outside in local hospital at 7 week of gestation, with normal cardiac activity and no other relevant findings. Anomaly scan was advised as part of normal routine evaluation of her pregnancy. Anomaly scan revealed duplicated lateral ventricle with dilated ventricular system and thinned-out brain parenchyma (Fig. 1A) suggestive of duplicated frontal lobe. The skull vault was deficient in the antero-superior aspect suggestive of acrania (Figs. 1A and 4). Two thalami were present which were not fused (Fig. 1A). Four eyes were noted in the face with 2 placed in the midline and 2 in the lateral aspect of the face (Figs. 1B and 4). There was 1 nose and 1 mouth in the face, suggesting only partial duplication of facial structure. Neck appeared normal

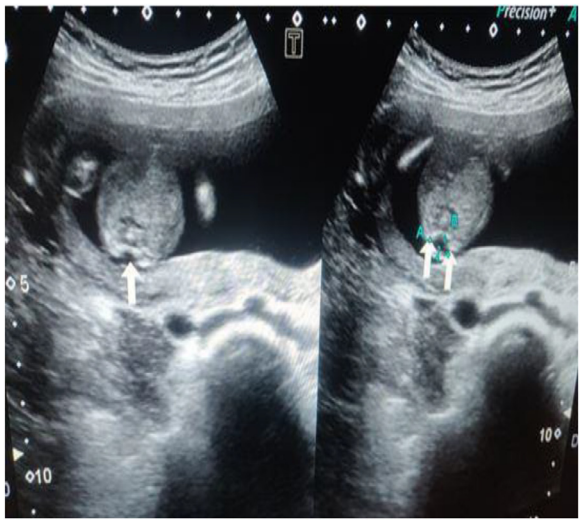

Fig. 2 – Axial B-mode ultrasound image of back showing presence of only 1 vertebral column with herniation of small amount of CSF and brain tissue via defect in posterior element in lower lumbar levels suggestive of small meningomyelocele (marked by white arrows).

with no evidence of cystic hygroma. There was 1 trunk with normal chest and abdominal structures. The posterior arch was deficit in lower lumbar region with herniation of small amount of brain tissue and cerebrospinal fluid suggestive of small meningomyelocele (Figs. 2 and 6). Both the feet were medially deviated suggestive of bilateral club foot (Figs. 3 and 5). Only 1 pair of upper limbs and lower limbs was present. There was no evidence of duplication of thoracic or abdominal structure however, dextrocardia was noted during scan. Single anterior placenta was noted with normal amniotic fluid volume with maximum vertical pockets of 3.6cm. Fetal cardiac activity

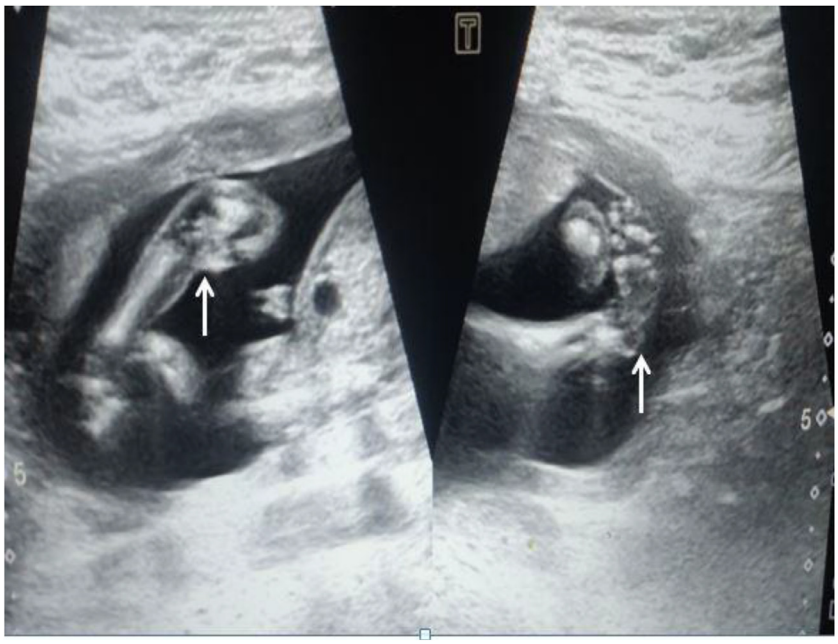

Fig. 3 – Oblique B-mode ultrasound image of bilateral lower limb showing both tibia and fibula in same image plane as of medially deviated foot—suggestive of bilateral club foots (marked by white arrows).

was regular with heart rate of 157bpm. So with constellation of findings of the duplicated frontal lobe with duplicated ventricular system, double thalami, partial absence of cranial vault, 4 eyes, small meningomyelocele, and bilateral club foot—the final diagnosis of diprosopus tetrophthalmus with acrania, spinal bifida and bilateral club foot was made. Couple counseling was provided regarding the poor prognosis of fetus due to various malformations, and termination of pregnancy was recommended. Post-abortus image of the fetus are shown in Figs. 4-6, which showed similar findings to the anomaly scan.

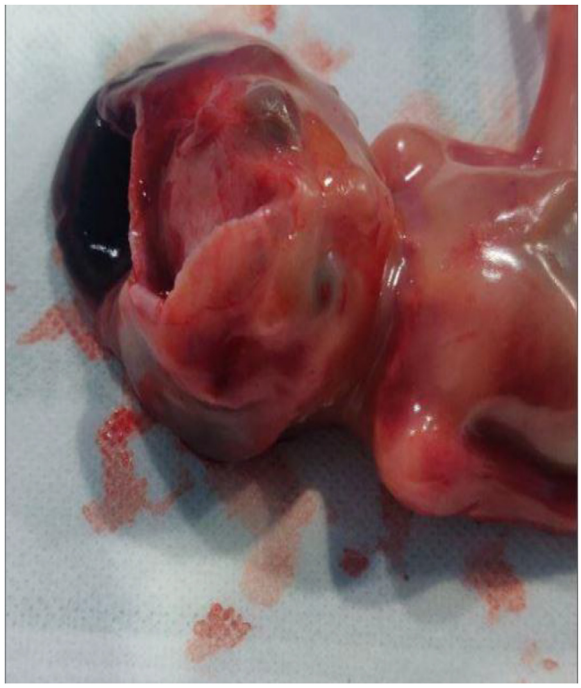

Fig. 4 – Postabortal image showing presence of 4 eyes with absence of skull vault. Note only 1 nose and mouth is present. Both upper limbs appear normal.

Discussion

Twin pregnancy can be monozygotic or dizygotic depending upon the number of initial zygotes formed during fertilization. Monozygotic twins can share placenta and amniotic cavity depending on time period of separation of the embryo resulting in dichorionic diamniotic, monochorionic diamniotic and monochorionic monoamniotic types of twin pregnancy [2]. Conjoint twins are rare types of twin that belongs to monozygotic monochorionic and monoamniotic category. Conjoint twins are defined according to parts of 2 twins fused with each other with thoracopagus (32.7%) being the most common and diprosopus (0.4%) being the least common variety [3]. Thoracopagus is defined as fusion in the thoracic region of twin whereas diprosopus is defined as presence of 2 faces in single head with variable degree of duplication of cranial and facial structure [1]. Prevalence of conjoint twin is 1 per 2800-200,000 deliveries with incidence of diprosopus being 1 in 180,000-15 million birth [3,1]. First presentation of diprosopus comes from an image with 2 heads found in the Neolithic sanctuary in Turkey, 6500 BC [4]. The exact etiology of Diprosopus is unknown, but there are various theories. One of which states that if there is defective oxidation between 15 and 25 days of implantation, there will be disruption in embryonic

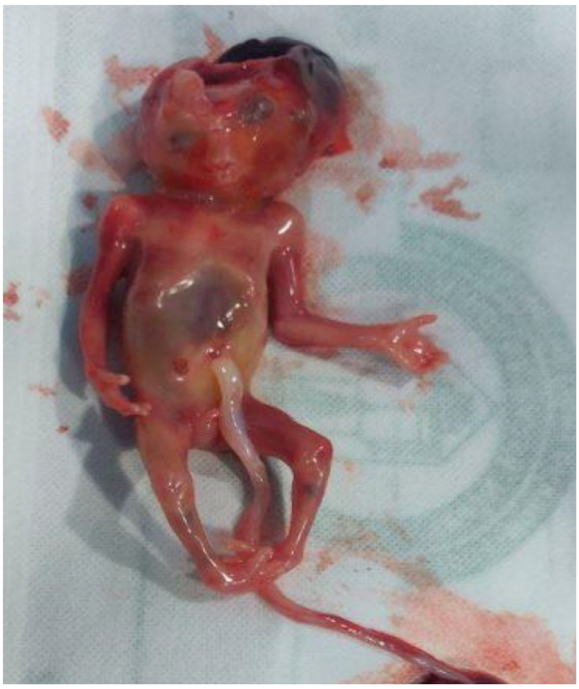

Fig. 5 – Postabortal image showing bilateral club foot. Note single nose and mouth with 1 trunk.

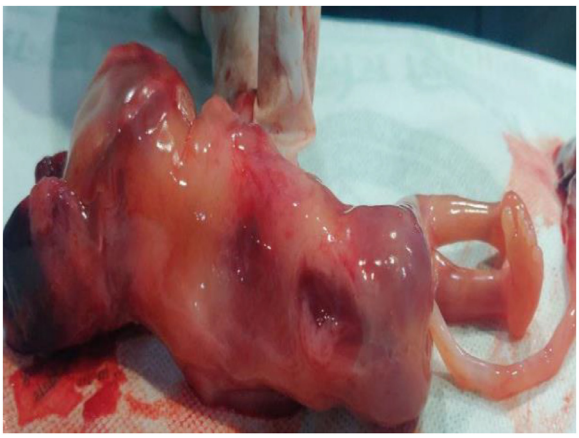

Fig. 6 – Postabortal image showing presence of small meningomyelocele in lower lumbar region.

development, with defective splitting leading to diprosopus formation [4]. The recent theory states that it results from 2 different notochords which were initially destined to become separate twins but were too close to each other and failed to develop independently [4]. Approximately half of the cases of diprosopus are associated with neural tube defects as in our case which showed acrania and small meningomyelocele [5]. Acrania in our case may represent spectrum of acrania-exencephaly-anencephaly where acrania leads to exposure of brain tissue to amniotic fluid causing its destruction and degeneration resulting in anencephaly. In diprosopus, there are 2 faces-like structures on single head and trunk with malformations ranging from partial duplication of some structures like the nose, mandible, mouth to all facial structures [1]. Our case showed duplication of eyes, frontal lobe of brain, ventricular system of brain and thalami. Other anomalies commonly associated with this condition are duplication of the spine, cardiac defects, club foot, and duplication of abdominal structures like stomach [1]. In 1993 Rating categorized diprosopus in 3 groups as follow [6].

Group I: Duplication takes place in upper half of face. There are 2 eyes and a nose to each face with common mouth.
Group II: Duplication takes place in lower half of face. There is single face with 2 mouths.
Group III: Duplication is symmetrical and may be parallel. There is duplication of nose and mouth but not of the eyes and ears.

Our case most likely belongs to group (I) however, in our case there was only 1 nose which is possible in diprosopus as there are variable degrees of duplication of facial structure. Anomaly scan plays a vital role in the diagnosis of this complex anomaly and expert radiologists are needed to confirm the diagnosis thus guiding the management of the patient. MRI of the brain combined with MR angiography would allow for a more thorough evaluation of this case by better delineating the brain anatomy and vascular duplication if any, which are lacking in our case report. However our case highlights the importance of anomaly scans during pregnancy in low-income countries like Nepal, where people of rural areas rarely do anomaly scans during their pregnancy. Outcome of diprosopus is very poor and many neonates with diprosopus are stillborn. Treatment option in survivors includes surgical repair in selected incomplete variety and management of complications arising out of other anomalies [7].

Conclusion

Even though diprosopus is rarest type of conjoint twin, treating physician and radiologist should be aware of this condition for proper and timely management of this condition.

Patient consent

Written informed consent for publication of this case report was obtained from coples.

REFERENCES

[1] D'Armiento M, Falleti J, Maria Maruotti G, Martinelli P. Diprosopus conjoined twins: radiologic, autoptic, and histologic study of a case. Fetal Pediatr Pathol 2010;29(6):431–8. doi:10.3109/15513815.2010.505628.
[2] Bidondo MP, Groisman B, Tardivo A, Tomasoni F, Tejeiro V, Camacho I, et al. Diprosopus: systematic review and report of two cases. Birt Defects Res A Clin Mol Teratol 2016;106(12):993–1007. doi:10.1002/bdra.23549.

[3] Hähnel S, Schramm P, Hassfeld S, Steiner HH, Seitz A. Craniofacial duplication (diprosopus): CT, MR imaging, and MR angiography findings— case report. Radiology 2003;226(1):210–13. doi:10.1148/radiol.2261011754.
[4] Olaya M, Rojas J, Franco JA. Craniofacial duplication: one of the rarest malformations in humans. Arch perinat med 2014;20(1):45–8.
[5] Rehder H, Kircher SG, Schoner K, Smogavec M, Behunova J, Ihm U, et al. Brain malformations in diprosopia observed in clinical cases, museum specimens and artistic representations. Orphanet J Rare Dis 2023;18(1):57. doi:10.1186/s13023-023-02617-5.
[6] Al Muti Zaitoun A, Change J, Booker M. Diprosopus (partially duplicated head) associated with anencephaly: a case report. Pathol - Res Pract 1999;195(1):45–50. doi:10.1016/S0344-0338(99)80094-6.
[7] Bhuyan M, Haque I. Diprosopus a rare craniofacial malformation. Asian J Neurosurg 2018;13(04):1257–9. doi:10.4103/ajns.AJNS_202_17.