Case Report

Rare Case of “Diprosopus Bicephalous Triophthalmus” and Review of Literature

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Introduction

Diprosopus or craniofacial duplication is one of the rarest congenital disorders of humans. The anomaly ranges from simple duplication of facial structure such as nose or eyes to complete duplication of the face. Systemic anomalies are common with complete duplication.[1,2] The prevalence of diprosopus is 2/100,000 births and constitutes 0.4% of all types of conjoined twins.[1] The anomaly is common in female than in male. Till date, only 36 such cases have been reported in the world literature with only a few case reports from India.[1-4] Here, we report a rare case of 24-week-old male fetus associated with anencephaly and craniospinal rachischisis.

Case Report

A 22-year-old female patient with gravida 2, para 0, living 0 and abortion 1 with a history of 24 weeks of amenorrhea, presented to casualty with bleeding per vagina and loss of fetal movements since 2 days. She was from a low socioeconomic background. She had not undergone any antenatal checkups. She had a previous history of abortion. The patient had a history of second-degree consanguineous marriage. Past and personal histories were not significant.

On general physical examination, it was observed that the patient was a middle-aged female, moderately built, and nourished. Pallor was present. Per abdomen examination revealed that the uterus was relaxed and uterine height was of 24 weeks. Fetal heart rate was absent. Per vaginal examination revealed partially closed os, admitting only tip of finger, and effacement was absent. Routine biochemical investigations were within normal limits. Hemoglobin level was 7.2 g/dL, hematocrit was 22%, and peripheral smear showed microcytic hypochromic and normocytic hypochromic red blood cells with normal white blood cells and platelets, giving a picture of dimorphic anemia. Coagulation profile was within normal limits.

Emergency antenatal ultrasound examination was carried out and it showed conjoint twins with loss of fetal cardiac activity. After obtaining informed consent from the patient and attenders, induction of labor was initiated with prostaglandins and the pregnancy was terminated by vaginal delivery. Dead male baby weighing 1800 g was delivered. After obtaining consent, the fetus was sent to pathology department for histopathological examination.

External examination revealed male fetus with head circumference of 30 cm (both the heads), chest circumference of 15 cm, abdominal circumference of 20 cm, and foot length of 7 cm. The fetus had complete duplication of

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cranial structure with three eyes. One eye of both the duplicated head was fused in the center. Two nose, two ears, two mouths, of which one lip showed cleft lip and palate and another showed only cleft lip, were observed. Fetus showed single short neck, thorax, and abdominal cavity. Both upper and lower limbs were normal.

Figure 1: Gross photograph of diprosopus, anterior (A) and lateral view (B), showing duplication of face with fusion in the center. Note the cleft lip and palate on right side. (C) Posterior view of fetus showing absence of brain and craniospinal rachischisis (white arrow). (D) (E) note the craniospinal rachischisis (red arrow). (F) CT image showing absence of occipital bone (white arrow head) along with craniospinal rachischisis (white arrow). (G) CT image highlighting craniospinal rachischisis (red arrow). (H) MRI images highlighting craniospinal rachischisis (black arrow). (I) MRI image showing two well-formed orbits along with fused central orbit (red arrow). (J) (K) (L) MRI images showing absence of brain and occipital bone (anencephaly along with other thoracic and pelvic viscera in situs solitus).
Brain was not developed and was replaced by grey brown tissue, showing features of anencephaly. Cervicothoracic and lumbar rachischisis were noted [Figure 1A-C].

Computerized tomography (CT) scan and magnetic resonance image (MRI) virtopsy were performed, which revealed duplication of face and confirmed the duplicated facial structures. Both the faces were fused in midline. Fusion of eye was observed in midline. The brain was hypoplastic. There were separate two sets of frontal, parietal, temporal, mandible for each face, which were fused in the center. Occipital bone was absent. One of the fetus had cleft lip and palate. A complete spina bifida with a cleft through the entire spine was observed, suggestive of rachischisis [Figure 1D-I]. The fetus had only one short neck, one thoracic, and abdomenopelvic cavity showing organs, which were in situ solitus [Figure 1H, J-L]. No anomaly was detected except for hypoplastic lungs. Autopsy was carried out, which confirmed findings of CT and MRI features. With all these above features, diagnosis of “diprosopus bicephalous triophthalmus” associated with anencephaly and craniospinal rachischisis was made.

**DISCUSSION**

The existence of conjoined twins is known since centuries, but they are reported in the literature since 1984. Ten percentage of monoamniotic, monochorionic, and monozygotic twins are conjoined. The most common of the conjoined twins is thoracopagus (32.7%), the rarest being diprosopus (0.4%). Diprosopus is a Greek word meaning “two-faced person.” The earliest description of diprosopus is credited to Ambroise Pare of sixteenth century. Approximately 36 cases of diprosopus are reported in world literature till date with a few cases from India.[1-4]

The exact etiology of the condition is unknown. Various mechanisms have been proposed but the most accepted one is due to the abnormality of sonic hedgehog genes and protein (Shh). Shh protein and corresponding genes are responsible for signaling and patterning of craniofacial structure. It also organizes the embryonic cells to specific areas, which later develops into specialized organs. In the brain, absence of Shh protein leads to holoprosencephaly and failure to move optic disc leads to cyclopia. If the activity of protein is increased, it leads to duplication of organs leading to diprosopus. Few authors also feel that the anomaly is due to the fusion of the parallel notochord in close proximation or fission of single notochord because of the abnormality of Dix homeobox gene. But till date, no genetic abnormality has been recorded with diprosopus.[1,2,5,6]

**Table 1: Systemic anomalies associated with diprosopus**

| System                      | Associated anomalies with diprosopus                                      |
|-----------------------------|--------------------------------------------------------------------------|
| Central nervous system      | Anencephaly, partial or complete duplication of cerebral hemisphere,     |
| Cardiovascular system       | spina bifida, craniorachischisis, duplication of cervical spine,         |
| Renal system                | arachnoid cyst, hypoplasia of temporal lobe                                |
| Gastrointestinal system     | VSD, over riding of aorta, hypoplastic aorta, dextrocardia               |
| Head and neck               | Dysplastic kidneys, hypoplasia of urinary bladder and ureter             |
| VSD = ventricular septal defect | Diaphragmatic hernia, imperforate anus                                     |

Diprosopus is associated with varied systemic anomalies. Anomalies are most common if there is complete duplication and less frequent in partial duplication. The reported congenital anomalies with diprosopus are shown in Table 1.[1-6]

The earliest clinical findings associated with diprosopus are polyhydramnios.[5] The disease can be diagnosed prenatally by four-dimensional ultrasound scanning, which will point out all the facial features. Associated congenital anomalies such as anencephaly, spina bifida, and other neural tube defects can be identified, but the facility is not available in rural areas. Estimation of serum alpha fetoprotein levels also helps in prenatal diagnosis.[1-5]

The prognosis of the condition depends on the degree of duplication, cases with complete duplication are stillborn (as observed in our case) and those with partial duplication involving one or more organs can be surgically corrected.[1,2]

**CONCLUSION**

A radiological and pathological finding of a rare case of “diprosopus bicephalous triophthalmus” is presented. Brief review of literature pertaining to possible etiology and associated congenital anomalies is discussed.

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**Conflicts of interest**

There are no conflicts of interest.

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