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CASE REPORT

Radiological features of pygopagus conjoined twins: A case report

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INTRODUCTION

Conjoined twins are rare developmental disorders of undetermined aetiology with varying degrees of prevalence from 1:50,000 to 1:200,000. A higher rate of stillbirth is observed among conjoined twins, which is predicted to be over 60%.^{1, 2, 3}

Conjoined twins are categorised by their primary connection site, such as thoracopagus, omphalopagus, pygopagus, ischiopagus, parapagus, craniopagus, cephalopagus, or rachipagus.⁴ Approximately 15% to 20% are classified as pygopagus, which are joined at the sacral region, sharing the spinal cord, genitourinary system, digestive system, and termination of the spine.⁵

Treating conjoined twins presents unique challenges to medical professionals. Accurate imaging, based on the connection site, is vital for surgical planning. Imaging must detect anatomical fusions, vascular anomalies, and related abnormalities to provide essential surgical and prognostic information, as each set of twins is unique.⁶

Here, we report the radiological findings of a unique case of pygopagus conjoined twins with fused spine, genitourinary, and other anatomical fusions.

CASE DESCRIPTION AND MANAGEMENT

Female conjoined twins of twenty-two days came from Kurigram (a northern district of Bangladesh) and were admitted to the Neurosurgery Department of Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka. The mother had no prior childbirth complications, chronic diseases, infections, radiation exposure, teratogenic drug use, or family history of similar and other congenital disorders. An ultrasound revealed the twin pregnancy, but the parents were not

LEARNING POINTS

- Conjoined twins challenge physicians of all disciplines, including radiologists, paediatric surgeons, neurosurgeons, plastic surgeons, and vascular surgeons.
- Detailed radiological evaluation of anatomical, vascular and other malformations in conjoined twins is crucial for proper surgical planning and prognosis.

informed that the twins were conjoined. The twins were delivered by caesarean section at 35 weeks, with a combined birth weight of 4.5 kilograms.

After admission, a medical team was formed consisting of a paediatric surgeon, neurosurgeon, anaesthesiologist, radiologist, plastic surgeon, and child nutritionist. A thorough clinical and radiological evaluation was conducted. On clinical examination, they were dorsally fused at the lumbosacral region sharing common perineum with a single anus and external genitalia with separate urethral meatus. The heart, lungs, and central nervous system were normal.

Afterwards, the babies were subjected to digital radiography, CT scans, and MRI for radiological evaluation. A CT scan revealed bony fusion of the spines at the level of the 5th sacral vertebra (S5) vertebra (FIGURE 1b) and fusion of soft tissues in the lower back (FIGURE 1a). The coccyx was not visible in either baby (FIGURE 1a). After administering per-rectal contrast, a separate rectum was seen for both babies, which fused at the S5 vertebra and resulted in a single anal canal (FIGURE 1c). MRI of the whole spine was also done, which revealed fusion of the dural sac at the level of the 2nd sacral vertebra (S2). Nerve roots were seen up to the fused portion of the dural sac (FIGURE 1d).

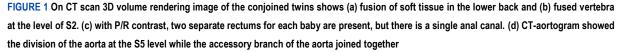
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Subsequently, a treatment plan was established. Each twin had two tissue expanders put over the chest and thigh region, gradually inflating over two and a half months to provide enough skin cover. To successfully separate the babies, a transverse loop colostomy was initially performed. Subsequently, a single-session separation surgery was completed without complications, separating the dural sac, sacral vertebra, and overlying soft tissue and skin. Separate anal canals were reconstructed for each baby. Currently, both babies are healthy, and their wounds are healing well.

DISCUSSION

Conjoined twins are a rare disorder that always shares the same gender. They are monozygotic, monoamniotic, and monochorionic and developed from a single fertilised ovum with a single placenta.Z.8 Like other conjoined twins, pygopagus twins are three times more prevalent in females, and females have a better survival rate.1.2 As the successful separation of conjoined twins is particularly difficult for paediatric surgeons, an imaging method to accurately detect anatomic fusion, vascular anomalies, and related abnormalities is critical

for proper surgical planning and prognostic information.^z

CT-scan combined with 3D reconstruction and standard bone algorithm provides good anatomical information and excellent assessment of bony fusion anomalies. MRI offers excellent visceral conjunction and overall anatomical depiction.⁶ As in this case, CT scans revealed a fusion of soft tissue and bone at the level of the S5 vertebra, which helped in deciding the size, shape and site of skin flapping. P/R contrast imaging showed separate rectums merging into a single anal canal, and MRI revealed dural sac fusion at the S2 vertebra. These findings helped in devising effective surgical procedures.

In summary, conjoined twins exhibit various foetal connections and anomalies. Evaluating conjoined twins requires specific imaging modalities based on the primary site of conjunction. While a general guideline exists, as each case is unique, precise imaging tailored to the specific twins and the requirements of the attending paediatric surgeons is necessary.

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Author contributions

Manuscript drafting and revising it critically: MS, MSS, SAA, PC, SS. Approval of the final version of the manuscript: MS, MSS, SAA, PC, SS. Guarantor of accuracy and integrity of the work: MS, MSS.

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Conflict of interest

We do not have any conflict of interest.

Ethical approval

Although ethical clearance was not sought, consent from the patient's guardian was obtained

Data availability statement

The authors confirm that the data supporting the findings of this study are shared upon request.

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