

CASE REPORT

Open Access



Emergency caesarean section in a patient with pygomeia: a curious tale of a misidentified “tail”—a case report

Reena^{1*}, Rahul Singh² and Ashutosh Vikram³

Abstract

Background: Polymelia or congenital duplication of the lower limb is a rare occurrence. This congenital duplication when occurring in the region of the buttocks is known as pygomeia. Its appearance can even mimic a human tail or pseudotail.

Case presentation: A 22-year-old primigravida posted for emergency caesarean section was found to have a tail-like structure attached to her lower back. Pre-operative imaging of that appendage to rule out spinal dysraphism was not done due to the need of emergency surgery and pregnant status of the patient. The patient received general anaesthesia for surgery, and in the follow-up, X-ray imaging was done which showed a hemipelvis and a femur bone in that appendage confirming the diagnosis of pygomeia.

Conclusions: In a rare occurrence of pygomeia, the exact nature can be identified by various imaging modalities. General anaesthesia was a safe choice in such a patient presenting for emergency caesarean section.

Keywords: Polymelia, Parasitic twin, Pygomeia, Congenital duplication of limb, Human tail, Pseudotail, Case report

Background

The incidence of conjoined twins has been reported to be 1.47 in 100,000 births worldwide (Mutchinick et al., 2011). Parasitic vestigial twins constitute only 10% of their population (Sharma et al., 2010). As the name suggests, the parasitic part thrives upon the autositic or living twin. They have further been classified on the basis of their most prominent site of fusion into the following: cephalopagus (head), thoracopagus (thorax), omphalopagus (umbilicus), rachipagus (spine), pygopagus (buttocks), ischiopagus (hip), craniopagus (cranium), and parapagus (side) (Spencer, 1996). Polymelia or congenital duplication of the lower limb is known as pygomeia when the parasitic nonviable twin is attached to the autosite via the buttocks, usually presenting as an incomplete supernumerary limb (Bastiani-Griffet et al., 1990). The appearance sometimes can be mistaken as a

human tail. We report the anaesthetic management of such a rare case undergoing emergency caesarean section for ante-partum haemorrhage (APH).

Case presentation

A 22-year-old full-term primigravida with active bleeding per vaginam was posted for emergency caesarean section. She had no significant past medical or surgical history. There were no previous ante-natal check-up records, and this was her first visit to a hospital, that too in an emergency with a history of fall from stairs resulting in ante-partum haemorrhage. She was pale, and her physical examination revealed a nearly midline tail-like structure attached predominantly to the left side of the buttock (Fig. 1). The length and width of that appendage were respectively 38.4 centimetres (cm) and 7.8 cm at the lower end. Her blood pressure was 112/64 mm Hg, and heart rate was 105 beats/minute. Her body weight was 84 kg, and height was 5 feet 3 inches. The rest of the physical examination did not reveal any other

* Correspondence: reena216@gmail.com

¹Institute of Medical Sciences, BHU, Varanasi 221005, India

Full list of author information is available at the end of the article



Fig. 1 Tail-like appendage at the back of patient

significant congenital anomalies or cardio-respiratory problems. Spinal examination also did not reveal any obvious abnormalities. Her haemoglobin was 8.5 g/dl. In view of active bleeding and impending foetal jeopardy, the patient was taken up for emergency caesarean section after ensuring availability of adequate blood products. Pre- and intraoperative patient's monitoring included electrocardiogram, pulse oximeter, non-invasive blood pressure, heart rate, and capnometry. She was unable to maintain supine position on the operation table due to the presence of tail as well as due to her pregnant status. A left lateral tilt of 30° was given to make her comfortable and also to displace gravid uterus. Two 18-gauge intravenous cannulas were inserted, and through each, 500 mL of lactated Ringer was administered fast.

Rapid sequence induction and intubation was planned in view of active bleeding, full stomach status, and most importantly presence of a tail which raised the suspicion of spinal dysraphism, which could not be ruled out pre-operatively by any imaging technique due to the pregnant status of the patient and emergent nature of surgery. The patient was pre-oxygenated with 100% oxygen for 5 min. Injection glycopyrrolate 0.2 milligram (mg) was given to reduce salivation and intravenous ondansetron 4 mg to prevent nausea/vomiting. Cricoid pressure was applied at the start of induction with intravenous thiopentone 250 mg followed by rocuronium 60 mg. The patient was intubated 90 s later with cuffed endotracheal

tube size 7, cuff inflated, and tube position was confirmed using capnography and bilateral equal air entry on chest auscultation. The tube was secured, connected to closed circuit, and intermittent positive pressure ventilation (IPPV) was started. Maintenance of anaesthesia was done with isoflurane in oxygen: nitrous oxide in 50:50 ratio and muscle relaxation maintained with intermittent top ups of intravenous rocuronium. Analgesia was maintained with injection fentanyl 100 µg, diclofenac 75 mg, and paracetamol 1 g infusion. The patient received one unit packed red blood cells intraoperatively. A healthy male baby with no obvious congenital anomalies was delivered with Apgar scores of 8 and 9 at 1 min and 5 min respectively. The patient was given injection oxytocin for adequate uterine contraction. Towards the end of the surgery, all inhalational anaesthetics were stopped, and neuromuscular reversal was done with neostigmine 2.5 mg and glycopyrrolate 0.2 mg after ensuring the patient's spontaneous respiratory efforts. The patient was extubated after thorough oropharyngeal suctioning and shifted to post-operative ward for overnight monitoring. Her vitals remained stable throughout the perioperative period. On the third post-operative day, the patient was counselled for getting the X-ray imaging of the "tail" done, which showed a complete femur bone (33.5 cm long) attached to a third hemipelvis (Figs. 2 and 3) and had no bony connection with the spine. Her electrocardiogram and two-dimensional echocardiogram both showed normal findings. Further imaging of the spine or



Fig. 2 Standard X-ray imaging showing AP view of appendage



Fig. 3 Standard X-ray imaging showing lateral view of appendage

the additional limb was not done as patient did not want to undergo any reconstructive surgery. The written informed consent to publish the article was obtained from the patient.

Discussion

In this peculiar case, diagnosis can fall into one of the following 3 categories:

1) Parasitic vestigial twin

The parasitic or heteropagus twin is a rare developmental anomaly, in which an extremely deformed vestigial remnant of the non-viable twin is dependent on its viable counterpart or “autosite” for its growth (Spencer, 2001). In literature review, exact data on the prevalence of parasitic twins is difficult to assess, and among these, data on pygopagus parasitic twin is even scarcer. Almost all of the reported cases are from the paediatric age group. Our case is unique in that way too, as it is the first case of pygopagus parasitic twin to be reported in an adult patient.

There are two popular theories describing the origin of heteropagus twinning: one is fission theory and another is fusion theory. Spencer et al. believed that an error during the blastogenesis causes incomplete “fission” of a single zygote while “fusion” theory proposes coalition of two distinct embryos prior to implantation (Spencer, 2000; Logroño et al., 1997). It is also postulated that subsequent intra-uterine selective ischemic insult results in demise and partial resorption of one of the twins which remains attached as a vestigial remnant to its surviving twin (Sharma et al., 2010; Spencer, 2000).

A number of terminologies have been used to describe this type of malformations including monstrosities, dipygus, pygomelia, lumbosacral duplication, or caudal duplication (Halidou-Doudou et al., 2008; Bajpai et al., 2004; Ngom et al., 2007). Complete dipygus is a complete lumbosacral duplication with two pelves, two pairs of buttocks, and four legs. Pygomelia is an incomplete dipygus resulting in development of one or more supernumerary members, which can be more or less complete and appended to the hip and may resemble a false tail (Bastiani-Griffet et al., 1990). In our patient too, a close inspection of the upper part of the supernumerary limb showed presence of two dimples corresponding to the extra buttocks, a finding similar to Nikiema et al. (Nikiéma et al., 2015).

2) Isolated congenital limb duplication

Isolated limb duplication is a rare congenital condition and has been interchangeably used for parasitic twinning also (Griffet et al., 2000). It is widely accepted that

adverse embryogenic influences are responsible for this kind of anomaly. Between the fourth and fifth weeks of embryonic development, limb differentiation occurs (Roberts & Tabin, 1994), and it follows a dorsal to ventral and proximal to distal pattern. Initially, 2 pairs of limb buds— anterior and posterior— protrude from both sides of the embryo and comprise cells of ectoderm and mesoderm. Their interaction is responsible for cell positioning and limb differentiation. The level and manifestation of limb deformity can thus be used to determine the approximate timing of the teratogenic event that occurred during limb development. Many factors have been implicated as contributing reasons for such malformations: trophic factors, traumatic, teratogenic substances, and genetic factors (Donnai & Winter, 1989). A comprehensive evaluation including physical, radiological, and electrophysiological examinations should be undertaken to obtain a detailed picture of the supernumerary limb and its relationship with surrounding structures.

3) Human tail or pseudotail

Another interesting diagnosis is “human tail” which has been classified as true tail or pseudo tail (Dao & Netsky, 1984). In the 5th–6th week of intrauterine life, human embryo has a tail with 10–12 vertebrae, which disappears by the 8th week of development (Fallon & Simandi, 1978). Persistence of a tail has often been associated with the occurrence of occult spinal dysraphism in the form of spina bifida, meningocele, lipomeningoceles, myelomeningocele, intraspinal lipoma, spinal cord tethering, or coccygeal vertebrae (Singh et al., 2008).

Regardless of the differential diagnosis, general anaesthesia was a safe choice in this case as it was an emergency case with patient presenting with ante-partum haemorrhage; also, there was not enough time to assess the nature of that “tail” pre-operatively. Since spinal dysraphism was a possibility in such a patient, spinal or epidural anaesthesia was not an option. Post-operatively, we counselled the patient that it is important to know the nature of the “tail” by radiographic imaging, which will be helpful if she plans to undergo reconstructive surgery in future. The X-ray imaging in our patient showed a third hemipelvis with a fully developed femur attached to it. Though there was no bony connection with the spine as seen in the X-ray image, spinal dysraphism in the form of tethered cord can only be ruled out by magnetic resonance imaging (MRI) scan.

Being born with an additional limb or tail is often associated with social stigma, shame, and fear of rejection. Delayed reporting and underreporting often occurs due to superstition and illiteracy in society. In our patient too, her parents did not seek medical attention after her

birth, and even she was living with it due to her lack of knowledge. The level of social stigma is such that she never visited hospitals for any ante-natal check-ups. Nevertheless, imaging is crucial for appropriate assessment and for planning subsequent management, should the patient decide for its surgical removal. Standard X-ray imaging is the main modality to assess bone formation and segmentation anomalies (Bastiani-Griffet et al., 1990). A thorough pre-operative systemic examination and investigations like electrocardiogram and two-dimensional echocardiogram are also necessary to exclude major cardiovascular malformations before the patient is scheduled for any elective surgery, since occasional occurrence of congenital heart defects such as ventricular septal defect, atrial septal defect, patent ductus arteriosus, and aortic valve dysplasia have been documented in association with these anomalies by several authors (Dejene et al., 2018; Zhao et al., 2006).

Conclusions

The presence of an additional limb or tail-like appendage in human has always been a matter of curiosity for medical practitioners. The exact nature can be identified by various imaging modalities. General anaesthesia was a safe choice in such a patient presenting for emergency caesarean section.

Abbreviations

APH: Ante-partum haemorrhage; IPPV: Intermittent positive pressure ventilation; MRI: Magnetic resonance imaging

Acknowledgements

None.

Authors' contributions

R. reviewed the available literature. RS and R. prepared the primary article. RS, R., and AV reviewed, edited, and approved the manuscript. The authors have read and approved the final manuscript.

Funding

None.

Availability of data and materials

The data and material of the case report are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Ethical approval is not required at our institution to publish an anonymous case report.

Consent for publication

The authors certify that they have obtained appropriate patient consent form. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Competing interests

The authors declare that they have no competing interests.

Author details

¹Institute of Medical Sciences, BHU, Varanasi 221005, India. ²Heritage Institute of Medical Sciences, NH-2, Bhadwar, Varanasi 221311, India. ³Narayan Medical College and Hospital, Sasaram, Bihar, India.

Received: 14 July 2021 Accepted: 20 February 2022

Published online: 07 March 2022

References

- Bajpai M, Das K, Gupta AK (2004) Caudal duplication syndrome: more evidence for theory of caudal twinning. *J Pediatr Surg* 39(2):223–225. <https://doi.org/10.1016/j.jpedsurg.2003.10.018>
- Bastiani-Griffet F, Bollini G, Berard J, Griffet J, Bouyala JM (1990) La pygome'lie: a propos de 2 cas et d'une revue de la litte' rature [Pygome'lia: about 2 cases and a literature review]. *Chir Pediatr*. 31:333–336
- Dao AH, Netsky MG (1984) Human tails and pseudotails. *Hum Pathol* 15:449–453. [https://doi.org/10.1016/s0046-8177\(84\)80079-9](https://doi.org/10.1016/s0046-8177(84)80079-9)
- Dejene B, Negash SA, Mammo TN, Getachew ATH, Derbew M (2018) Heteropagus (parasitic) twins. *J Pediatr Surg Case Rep* 37:44–49. <https://doi.org/10.1016/j.jeps.2018.07.019>
- Donnai D, Winter RM (1989) Disorganisation: a model for 'early amnion rupture'? *J Med Genet* 26:421–425. <https://doi.org/10.1136/jmg.26.7.421>
- Fallon JF, Simandi BK (1978) Evidence of a role for cell death in the disappearance of the embryonic human tail. *Am J Anat* 152:111–129. <https://doi.org/10.1002/aja.1001520108>
- Griffet J, Bastiani-Griffet F, Jund S, Moreigne M, Zabjek KF (2000) Duplication of the leg—renal agenesis: congenital malformation syndrome. *J Pediatr Orthop B* 9:306–308. <https://doi.org/10.1097/01202412-200010000-00015>
- Halidou-Doudou M, Hallarou M, Tambary MD, Harakoye A, Manzo ML, Adamou H (2008) Pygome'lus: a surgical case report from Nigeria. *Arch Pediatr* 15:1426–1429. <https://doi.org/10.1016/j.arcped.2008.06.017>
- Logroño R, Garcia-Lithgow C, Harris C, Kent M, Meisner L (1997) Heteropagus conjoined twins due to fusion of two embryos: report and review. *Am J Med Genet* 73:239–242. [https://doi.org/10.1002/\(SICI\)1096-8628\(19971219\)73:3<239::AID-AJMG1>3.0.CO;2-N](https://doi.org/10.1002/(SICI)1096-8628(19971219)73:3<239::AID-AJMG1>3.0.CO;2-N)
- Mutchinick OM, Luna-Muñoz L, Amar E, Bakker MK, Clementi M, Cocchi G et al (2011) Conjoined twins: a worldwide collaborative epidemiological study of the International Clearinghouse for Birth Defects Surveillance and Research. *Am J Med Genet C Semin Med Genet* 157C:274–287. <https://doi.org/10.1002/ajmg.c.30321>
- Ngom G, Sankale AA, Fall M, Die'me' C. (2007) A case of dipygus treated surgically. *Internet J Orthop Surg* 6(2)
- Nikiéma Z, Kouamé N, Dakouré PWH, Traore C, Rabiou R (2015) Dipygus: Computed tomography findings and management. *Diagn Interv Imaging* 96:1219–1221. <https://doi.org/10.1016/j.diii.2013.04.018>
- Roberts DJ, Tabin C (1994) The genetics of human limb development. *Am J Hum Genet* 55:1–6
- Sharma G, Mobin SSN, Lypka M, Urata M (2010) Heteropagus (parasitic) twins: a review. *J Pediatr Surg* 45(12):2454–2463. <https://doi.org/10.1016/j.jpedsurg.2010.07.002>
- Singh DK, Kumar B, Sinha VD, Bagaria HR (2008) The human tail: Rare lesion with occult spinal dysraphism- a case report. *J Pediatr Surg* 43:e41–e43. <https://doi.org/10.1016/j.jpedsurg.2008.04.030>
- Spencer R (1996) Anatomic description of conjoined twins: a plea for standardized terminology. *J Pediatr Surg*. 31(7):941–944. [https://doi.org/10.1016/s0022-3468\(96\)90417-0](https://doi.org/10.1016/s0022-3468(96)90417-0)
- Spencer R (2000) Theoretical and analytical embryology of conjoined twins: part I: Embryogenesis. *Clin Anat* 13:36–53. [https://doi.org/10.1002/\(SICI\)1098-2353\(2000\)13:1<36::AID-CA5>3.0.CO;2-3](https://doi.org/10.1002/(SICI)1098-2353(2000)13:1<36::AID-CA5>3.0.CO;2-3)
- Spencer R (2001) Parasitic conjoined twins: external, internal (fetuses in fetu and teratomas), and detached (acardiacs). *Clin Anat* 14:428–444. <https://doi.org/10.1002/ca.1079>
- Zhao L, Li MQ, Sun XT, Ma ZS, Guo J, Huang YT (2006) Congenital lumbosacral limb duplication: a case report. *J Orthop Surg* 14(2):187–191. <https://doi.org/10.1177/230949900601400216>

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Submit your manuscript to a SpringerOpen® journal and benefit from:

- Convenient online submission
- Rigorous peer review
- Open access: articles freely available online
- High visibility within the field
- Retaining the copyright to your article

Submit your next manuscript at ► [springeropen.com](https://www.springeropen.com)