

Contents lists available at [ScienceDirect](http://ScienceDirect.com)

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpascasereports.com

Oral fetus-in-fetu: A case report



Nurudeen Toyin Abdulraheem*, Abdurashed A. Nasir, Lukman O. Abdur-Rahman, Oluwaseun R. Akanbi, Moses O. Olanrewaju, Muslimat A. Alada, Olanrewaju O. Oyedepo, Temitope Obasa, James O. Adeniran

Division of Paediatric Surgery, Department of Surgery, University of Ilorin, University of Ilorin Teaching Hospital, Ilorin, Nigeria

ARTICLE INFO

Article history:

Received 28 January 2015

Received in revised form

3 February 2015

Accepted 20 February 2015

Key words:

Fetus-in-fetu

Oral monster

Teratoma

Monozygotic twins

ABSTRACT

Fetus-in-fetu is a rare condition, less than 200 cases have been reported. Its embryopathogenesis is linked to a monozygotic, diamniotic parasitic twin. The presence of a calcified vertebral column and other body parts are key to the diagnosis, and differentiate it from a teratoma. We report a case of a neonate who was admitted immediately after delivery by Caesarian section following a prolonged obstructed labor caused by a huge mass projecting from the hard palate. The mass had identifiable malformed body parts but was anencephalic. Intraoperative findings were a short stalk and cleft of the soft palate. He had excision of the mass and did well post operatively. Persistent mouth breathing and difficult nasal cannulation lead to request for post operative magnetic resonance imaging which showed patent nostrils and absent residual mass. Prompt and skillful anesthesia and surgical intervention assisted in the survival of this patient.

© 2015 Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Fetus-in-fetu (FIF), a term first used by Meckel in the late 18th century, defines an abnormal embryogenesis of twinning [1–4]. An abnormal monozygotic twin develops inside the body of its host twin [2]. They are usually encapsulated, pedunculated vertebrate tumor, yet anencephalic [5]. The overall incidence is 1 in 500,000 [2]. Less than 200 cases are cited in the literature [3,5]. Many of these cases fail to fulfill Lord's criteria of diagnosis i.e. presence of calcified vertebrae and any other body parts [5]. Most common locations are retroperitoneal (80%), Skull (8%) and sacrococcygeal region (8%) [2]. We report a case of oral fetus-in-fetu (FIF) in a female neonate.

1. Case presentation

A female new born was admitted from the Obstetrics theater on account of a large outgrowth from the mouth noticed immediately after delivery. She was delivered at term (41wks by estimation) via emergency Caesarean section, on account of prolonged obstructed labor previously attended at home, to a 40 yr old P6⁺ (6 alive) housewife. Apgar scores were 7 at 1 min and 9 at 5 min. Mother was

unbooked and had no antenatal care during pregnancy. However, she did an obstetric ultrasound scan at around 24 weeks gestational age. Abnormality was detected in the fetus for which she was offered termination of Pregnancy. She (in agreement with her husband) declined for religious reasons and never sought further medical attention. She took herbal concoctions but had no exposure to ionizing radiation. No rash or fever in pregnancy. No history suggestive of consanguinity.

On examination, she was conscious; birth weight was 4.5 kg (with the mass). There was a large mass emanating from the mouth attached to the roof of the mouth by a fleshy stalk. Most of the mass was covered by skin, with multiple ulcerated points and deformed limbs (Fig. 1). The mass measured 20 cm × 20 cm × 10 cm and had mixed consistency. No other anomaly was detected. Working diagnosis was a palatal fetus-in-fetu.

The airway was secured by skillful anesthetic technique following a difficult endotracheal intubation. The mass was removed by ligating the stalk. Intraoperative findings included a grotesque mass with poorly formed feet, back and exposed intestines with most parts covered by skin attached to the hard palate by a wide stalk with a cleft soft palate. The facial structures were essentially normal.

She was noticed to be mouth-breathing on the first post-operative day. A nasogastric tube could only be passed through the right nostril. A left choanal atresia was then suspected necessitating

* Corresponding author. Department of Surgery, University of Ilorin Teaching Hospital, PMB 1459, Ilorin, Nigeria. Tel.: +2348035532580.

E-mail address: free2nta@yahoo.com (N.T. Abdulraheem).



Fig. 1. The patient showing a large mass emanating from the roof of the mouth with deformed limbs.

a review by the otorhinolaryngologists, who ordered a cranial MRI that showed no residual tumor at the roof of the mouth and patent nostrils. Quiescent left parieto-occipital extradural and subgaleal hematomas were also revealed.

She subsequently did well and was discharged 10 days post operatively. Histopathology result confirms a matured cystic teratoma. She remains well six months after operation (Fig. 2).



Fig. 2. The patient at 6 months of age.

2. Discussion

Fetus-in-fetu (FIF) describes an abnormal monozygotic twin which develops inside the body of its host [2]. Prashantha et al. reported a case they described as fetus-ex-fetu (FEXF) in which a malformed fetus was attached to a live fetus externally [6]. The case we report here is similar to the one described by Prashantha et al. Although, the mass was connected to our patient at the roof of the mouth, it was entirely outside the body.

Many of the reviewed literature described a monozygotic, diamniotic twinning that went awry because of unequal division of totipotent inner cell mass [2,5–8]. This is supported by a study that showed that intra-abdominal gonads, where present in the tumor, are similar histologically to the sex of the host [1]. However, this is not so in the case reported by Abdur-Rahman et al. where a mass with a rudimentary phallus and descended testicles were found on a female host [1]. This made them to propose that dizygotic parasitic fetiform twin could exist and when it does occur, may probably be superficially or externally located [1]. Our case may be this variant and genetic evaluation would be important. Furthermore, monozygotic twin theory may not be sufficient to explain the occurrence of multiple FIFs and association of FIF with teratoma as reported by Houshang [4]. Postulation therefore, includes defective implantation of blastocyst as the underlying mechanism [9].

Age at presentation varies from prenatal to as high as 47 years [1–11], however, less than 10% of cases present at age 10 years and above [1]. The externally located ones, like our case present immediately after birth, because of the grotesque appearance of the mass. The obscure ones e.g. the retroperitoneal type (which incidentally is the commonest) may tarry a while before presenting. Less than 10 cases of FIF have been reported from Africa [1].

Location of FIF varies widely. The most common site is retroperitoneal (80%), followed by skull (8%) and sacrococcygeal region (8%) [2]. Other less common sites include pelvis, scrotum and mediastinum [2]. The skull variety may be intracranial [2], attached to the face [6], or to the oral cavity like the case we present.

Symptoms of FIF typically relates to its mass effect. Abdominal FIF presents with abdominal distension [4,8–10], jaundice, intestinal and urinary obstruction [4]. Oral cavity FIF usually present with mass protruding through the mouth and cleft palate. Cranial FIF would give rise to neurological problems [4]. The case we present has a cleft of the soft palate. Prenatal diagnosis of FIF is on the increase with the aid of obstetric ultrasound scans and magnetic resonance imaging (MRI) in some cases [7,9]. However, misdiagnosis as a meconium cyst was reported and attributed to lack of consideration of FIF as a differential diagnosis [10]. Prenatal diagnosis offers the opportunity of expediting delivery and prompt treatment [2], and affords avoidance of preventable fetal loss [9].

Even with prenatal diagnosis, post natal evaluation with Ultrasound, MRI and/or Computerized Tomography (CT) may still be needed to confirm diagnosis [2,7,10]. Preoperative imaging was not done in this case for logistic reasons. Previous reports have emphasized the need for further preoperative evaluation of the mass, not for diagnosis, but, to know the extent of the disease and the nature of the attachment [1,6].

Serological evaluation may also be necessary as there have been reports of elevated α - Fetoprotein (AFP) [7,10] and β Human Chorionic Gonadotrophin (β – HCG) [7] both of which became normal after excision [7,10].

Anesthesia management in this index case was particularly challenging, same with a similar case reported by Mallick et al. [6]. Otorhinolaryngologist should always be on the standby to salvage and manage a difficult airway. Like in the case we are reporting, the mass was typically suspended by a pedicle [5,6]. However, intracavitary varieties may lack major vascular connections [2,8] and

derive their vascular supply from broad based surrounding tissue, hence dense adherence to these tissues should be expected [7]. When they are typically well encapsulated [2–5,7,8,10], a complete excision of the mass with its surrounding membrane is the treatment of choice, failing which malignant recurrence is a possibility [4].

Prognosis is very good as recurrence is rare [1,3–10] and more favorable than cystic teratomas [4]. Close clinical, radiological and serological follow up is recommended especially if immature elements are detected in the mass [4]. Immediate post operative radiological evaluation is also recommended especially for patients that never had such investigation prior to surgery. This will afford surgeons the opportunity to detect early, the presence of residual mass or other anomalies, and plan subsequent management.

3. Conclusion

Fetus-in-fetu is a very rare congenital anomaly. Less than 10 cases have been reported from Africa. Prompt and skillful anesthesia and surgical intervention assisted in the survival of this patient. Correct clinical diagnosis of fetus-in-fetu can be made from appearance and the components of the mass. Further evaluation especially radiological, may be indicated to identify the extent of the disease.

References

- [1] Abdur-Rahman LO, Abdul-Kadir AY, Rahman AG. Fetus-in-fetu in a 6-month-old. *Afr J Paediatr Surg* 2008;5:96–8.
- [2] Maryńczak L, Adamek D, Drabik G, Lankosz-lauterbach J. Fetus in fetu: a medical curiosity—considerations based upon an intracranially located case. *Childs Nerv Syst* 2014;30:357–60.
- [3] Rahman GA, AbdulKadir YA, Abdur-Rahman LO. Fetus-in-fetu: a review article. *Eur J Sci Res* 2007;18:663–73.
- [4] Pourang H, Sarmadi S, Mireskandari S-M, Soleimani M, Mollaeian M, Alizadeh H, et al. Twin fetus in fetu with immature teratoma: a case report and review of the literature. *Arch Iran Med* 2009;12:507–10.
- [5] Dutta HK, Thomas JK, Sahewala NK, Patgiri DK. Fetus in fetu in a neonate: report of a case. *Surg Today* 2013;43:547–9.
- [6] Mallick PN, Upadhaya SP, Das AK, Singh RKA. Anaesthesia management in foetus-ex-fetu. *Indian J Anaesth* 2013;57:2013–5.
- [7] Chua JHY, Chui CH, Sai Prasad TR, Jacobsen AS, Meenakshi A, Hwang WS. Fetus-in-fetu in the pelvis: report of a case and literature review. *Ann Acad Med Singapore* 2005;34:646–9.
- [8] Arlikar JD, Mane SB, Dhende NP, Sanghavi Y, Valand AG, Butale PR. Fetus in fetu: two case reports and review of literature. *Pediatr Surg Int* 2009;25:289–92.
- [9] Has R, Kalelioglu IH, Esmer AC, Demirbas R, Yuksel A, Yavuz E. Prenatal sonographic diagnosis of fetus in fetu. *J Ultrasound Med* 2013;32:2212–4.
- [10] Kim YJ, Sohn SH, Lee Y, Sohn JA, Hee E, Kim EK, et al. Misdiagnosis of fetus-in-fetu as meconium peritonitis. *Korean J Pediatr* 2011;54:133–6.
- [11] Khan N, Klimo Jr P, Harreld J, Armstrong GT, Michael 2nd LM. Mature teratoma of the petrous bone with extension into the cerebellopontine angle: case report. *J Neurol Surg Rep* 2013;74:96–100.