In airway malacias the contour of airways is maintained by the bronchial smooth muscle tone [5]. Infants with congenital airway malacias, presenting with wheeze may not improve with beta agonist nebulisation because in such lesions, beta agonists by reducing the muscle tone can aggravate the pathology. The child's improvement of respiratory problems with discontinuing of beta-agonist and continuation of semisynthetic anticholinergics (ipratropium) nebulization substantiates this. Though surgical interventions like aortopexy may be required in severe cases, majority of milder forms of airway malacias invariably improve with regular chest physiotherapy and postural drainage [1].

This case highlights the important principle that young infants presenting with recurrent wheeze and respiratory distress with poor response to bronchodilators may be advised flexible bronchoscopy as the initial investigation of choice for early diagnosis and effective management of airway malacias.

Contributors: DV has conceptualized the study and contributed to writing the manuscript. SK was involved in data analysis, outcome assessment, literature review and writing the manuscript. VEV was involved in critical analysis of the manuscript.
Funding: None.
Competing interests: None stated.

REFERENCES

1. Carden KA. Tracheomalacia and tracheobronchomalacia in children and adults–an in-depth review. Chest. 2005;127:984-1005.
2. Austin J, Ali T. Tracheomalacia and bronchomalacia in children: Pathophysiology, assessment, treatment and anesthesia management. Pediatr Anesth. 2003;13:3-11.
3. Mayer S, Laura N, Gerald Z, Joseph A. Pediatric intrathoracic large airway obstruction: Diagnostic and therapeutic considerations. Pediatric Emergency Care. 1994;10:313-2.
4. Yalç1n E, Do ru D, Özçelik U, Kiper N. Airway malacia disorders in children. Chest. 2006;130:304.
5. Panitch HB, Keklikian EN, Motley RA, Wolfson MR, Schidlow DV. Effect of altering smooth muscle tone on maximal expiratory flows in patients with tracheomalacia. Pediatr Pulmonol. 1990;9:170-6.

Double Fetus In Fetu

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Received: April 12, 2010;
Initial Review: May 07, 2010;
Accepted: August 3, 2010.

Fetus in fetu (FIF) is an extremely rare cause of infantile abdominal mass where a rudimentary, malformed monozygotic-diamniotic twin grows inside the other twin. We describe a male infant with double or twin fetuses in fetu. The diagnosis was made on a computerized tomography (CT) scan of the abdomen and confirmed on surgery. Surgical excision was done and the baby did well post operatively.

Key words: *Computerized tomography, Double fetuses in fetu, Teratoma.*

Fetus in Fetu (FIF) is an entity where one vertebrate underdeveloped twin develops inside the other normal host twin. Till date only about 100 cases have been reported. Most of the case reports describe a single FIF. We report a case of double FIF in a six-week-old infant.

CASE REPORT

A 6 weeks old infant presented to us with an abdominal lump and vomiting. The infant was born full term by normal vaginal delivery with a birthweight of 2500g. The baby was well till 2 weeks of life when the mother noticed an abdominal lump, which gradually increased in size leading to abdominal distention. The infant also started vomiting after feeds but continued to pass normal stools. On presentation to our hospital, the child weighed 3500g and his vitals were maintained. On abdominal examination there was distension and there was a well defined firm, round, non-tender mass in the right upper abdomen, occupying the right hypochondrium and lumbar region

extending a little into the iliac fossa. The bowel sounds were normal. CT abdomen revealed a heterogenous well encapsulated mass in the upper abdomen displacing the liver, bowel and stomach anteriorly (***Fig.* 1**). The mass was divided by thin septa into two compartments. Both the compartments contained two separate soft tissue shadows with separate sets of ossific elements resembling a vertebral column with a rudimentary rib cage. A provisional diagnosis of double or twin fetus in fetu was made and the infant was taken up for surgery. During surgery, a large cystic retroperitoneal mass was visible pushing the liver upwards and the rest of the abdominal contents to the left. The liver was mobilized and after further dissection and removing the peritoneum, the cystic mass was exposed. An attempt was made to remove the whole cystic mass as such but to prevent injury to the portal structures the capsule had to be incised. This revealed two fetuses lying in a pool of amniotic fluid. The blood supply to the sac was derived from the abdominal aorta of the baby and the venous drainage was to the inferior vena cava. The mass was resected. The post-operative period went uneventful and the infant was discharged on the 10th postoperative day. The child is doing well on follow up.

On examination, one of the fetuses weighed 250g (fetus A) and the other one weighed 180g (fetus B) (***Fig.* 2**). On gross examination fetus A showed scalp hair, optic pits, rudimentary auricles, primitive fore and hind appendages. There was no digit formation. On dissection a complete vertebral column was seen. Stomach and small intestine were developed. Rudimentary liver and left kidney was visible. The cranial cavity was filled with clear fluid and was devoid of any brain tissue. A malformed heart with no defined chambers was seen lying in the cervical region. Fetus B was less developed than its twin. It also had scalp hair but only a single rudimentary forelimb and the caudal end was not differentiated. On dissection the vertebral column was seen along with the stomach. No other abdominal organs were differentiated.

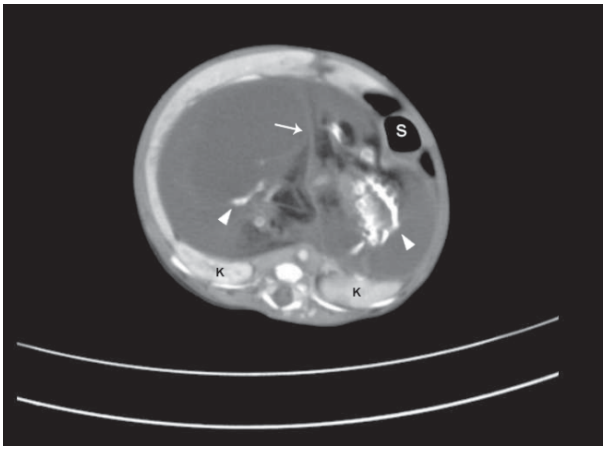

FIG. 1 *(a) Axial contrast enhanced CT showing a cystic mass divided by a septa (arrow) containing two sets of ossific elements (arrow heads). The mass has pushed the kideys (K) posteriorly and the stomach (S) anteriorly.*

DISCUSSION

Fetus in fetu (FIF) is a very rare cause of an abdominal mass in an infant. Multiple fetuses in fetu are still rarer [1-3]. FIF has been believed to arise from inclusion of one monozygotic- diamniotic twin into the other twin. There have been reports of FIF being detected as early as in the newborn period or even as late as adulthood [4]. Few cases of FIF have been detected antenatally as a cystic intra-abdominal mass growing inside the fetus [5]. The most common site is the retroperitoneum but FIF have been reported at various sites right from the cranial cavity to the scrotal sac [6,7]. The monozygosity of the twins can be confirmed by presence of identical sex karyotype, histocompatibility types and blood groups.

There has been a controversy regarding the differentiation between a teratoma and FIF. Some even consider them to be the two ends of a spectrum, FIF being a highly organized form of a teratoma [8]. Differentiation criteria between the two were suggested [9]. Teratomas are considered to arise from pluripotent cells but do not demonstrate vertebrae or systemic organogenesis. The commonest site for a teratoma is the sacrococcygeal region and they have a definite malignant potential. In contrast, the FIF are usually retroperitoneal, have a vertebral skeleton, are benign and usually have variably differentiated organ system and limbs.

In our patient, the diagnosis of FIF was suggested by presence of a vertebral column in the mass on an

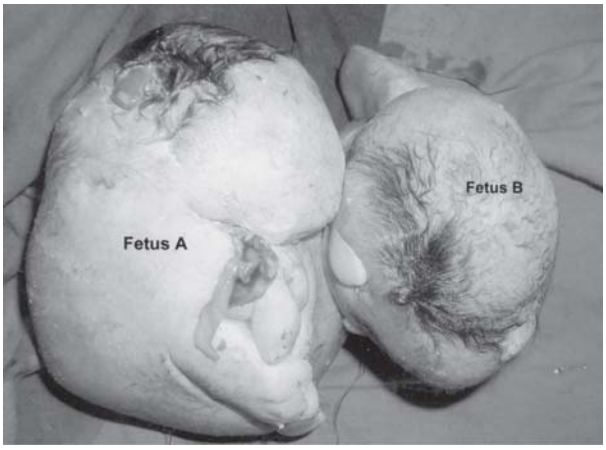

FIG. 2 *Photograph of the twin fetuses in fetu.*

abdominal *X*-ray. CT imaging gives a more accurate diagnosis and defines the relation of the mass with the other intra-abdominal structures [10]. The twin fetuses in fetu in our case fulfilled the criteria for being FIF and not a teratoma. Identification of a vertebral column indicates development of the parasite twin at least up to the stage of notochord from which the vertebral column arises. Symptoms of FIF are primarily due to its mass effect such as abdominal distention, feeding difficulty, emesis, jaundice or pressure effects on the renal or gastrointestinal system. Complete excision as was done in our patient ensures definitive cure .

Contributions: RR and DA were involved in case management and writing the manuscript. DKS was involved in drafting the manuscript and searching the literature. PK provided expert radiological opinion and critically reviewed the manuscript.
Funding: None.
Competing interests: None stated.

REFERENCES

1. Pourang H, Sarmadi S, Mireskandari SM, Soleimani M, Mollaeian M, Alizadeh H, *et al*. Twin fetus in fetu with immature teratoma: A case report and review of the literature. Arch Iranian Med. 2009;12:507-10.
2. Daga BV, Chaudhary VA, Ingle AS, Dhamangaokar VB, Jadhav DP, Kulkarni PA. Double fetus-in-fetu: CT scan diagnosis in an adult. Indian J Radiol Imaging. 2009;19:216-8.
3. Gangopadhyay AN, Srivastava A, Srivastava P, Gupta DK, Sharma SP, Kumar V. Twin fetus in fetu in a child: a case report and review of the literature. J Med Case Reports. 2010;4:96.
4. Dagradi AD, Mangiante GL, Serio GE, Musajo FG, Menestrina FV. Fetus-in-fetu removal in a 47-year-old man. Surgery. 1992;112:598-602.
5. Mills P, Bornick PW, Morales WJ. Ultrasound prenatal diagnosis of fetus in fetu. Ultrasound Obstet Gynaecol. 2001;18:69-71.
6. Afshar F, King TT, Berry CL. Intraventricular fetus-in-fetu. J Neurosurg. 1982;56:845-9.
7. Kakizoe T, Tahara M. Fetus in fetu located in the scrotal sac of the newborn infant. J Urol. 1972;107:506-8.
8. Potter EL. Pathology of the fetus and the newborn. *In:* Potter EL, eds. Pathology of the fetus and newborn. 2nd ed. Chicago, III: Year book, 1961. p. 183-7.
9. Willis RA. The borderland of embryology and pathology. Bull N Y Acad Med. 1950;26:440-60.
10. Iyer KV, Vinayak K, Haller JO, Maximin S, Barrerras J, Velchek F. Multiple fetuses in fetu: Imaging findings. Pediatr Radiol. 2003;33:53-5.