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Case Report

Anesthesia for Fetus in Fetu with Difficult Airway, Skeletal Dysplasia and Hyperparathyroidism

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Abstract

Fetus-in-fetu (FIF) is a rare condition in which malformed parasitic twin grows inside the body of its twin and usually presents as an abdominal mass. We are reporting a case of a neonate who presented with abdominal mass affecting the breathing with difficult airway; FIF was suggestive after further radiological studies. Intra-operatively, retroperitoneal the mass was identified in the sub hepatic region and successfully resected. Post operatively patient recovered well and discharged in good condition.

Keywords: *Fetus in Fetu; Difficult Airway; Teratoma; Retroperitoneal Neoplasms*

Background

Fetus in fetu (FIF) is very rare anomaly, which has been reported several times in the last few years. In FIF there is a parasitic twin grows inside the body of its twin, most commonly is found in the abdominal cavity. However, there were several cases where FIF was found intra cranial, mediastinal or scrotal. FIF is differentiated from teratoma by the presence of vertebral column often with an appropriate arrangement of other organs or limbs around it. Therefore, we are reporting a case of neonate with FIF as abdominal mass affecting the airway; to enrich the literature with such rare condition.

Case Report

Full term neonate admitted to NICU with dysmorphic features of Micrognathia and intra-abdominal mass affecting the breathing, which was first detected at 34 weeks of gestation during an antenatal ultrasound examination of 29-year-old Saudi female medically free, gravida 5 para 4. Ultrasound showed a vascular intra-abdominal mass occupying part of liver space which is adjacent to the bowel and right kidney, also all long bones were short and right radius, tibia and fibula were bowed with suspicion of fracture, chest was narrow and small. At 39 weeks of gestation mother underwent spontaneous vaginal delivery and delivered a baby girl with of 3.030-kg birth weight and Apgar score of 5 in first minute and 8 in eighth minute. Clinically patient was jaundiced with distended abdomen affecting breathing and palpable mass over the right upper quadrant.

Result of the lab investigations revealed high total and direct bilirubin in a pattern of cholestatic jaundice.

Alfa feto protein (AFP), beta human chronic gonadotrophin (β-HCG) and parathyroid hormone (PTH) levels were reported at 31986 µg/L,8 IU/L, and 2300 pg/ml respectively.

Moreover, genetic study showed negative results for aneuploidy 21, 18 and 13. Diffuse osteopenia, multiple healing fractures and evidence of cortical erosions were shown via Postnatal Skeletal survey, mainly, Metabolic Bone Disease was suggested based on the features of abnormality of upper and lower bones, subperiosteal resorption and hyperparathyroidism (Figure 1).

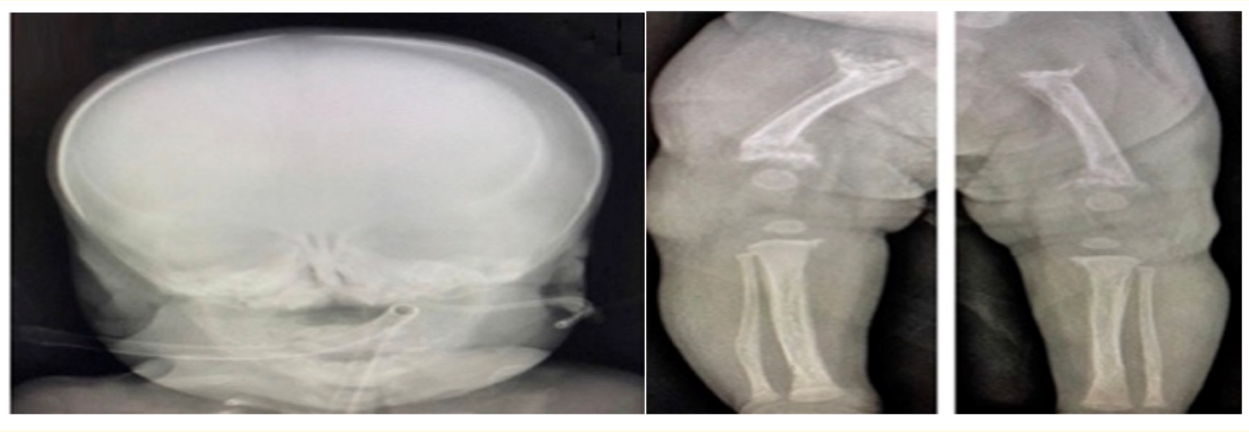

Figure 1: *Diffuse osteopenia.*

A large heterogeneous intrabdominal mass was witnessed via abdominal ultrasound which was located in the right upper quadrant closely pushing left lobe of the liver with intralesional cystic changes and specks of calcification (Figure 2). To highlight that mesenteric teratoma or liver primary lesion like hepatoblastoma might be considered the differentials.

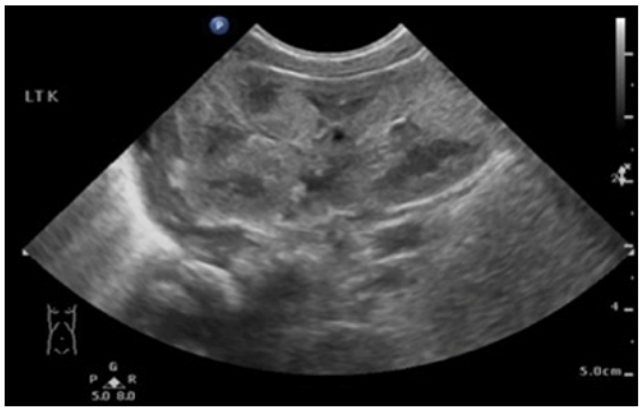

Figure 2: *Abdominal ultrasound showing the mass.*

A computed tomography scan CT of the patient's abdomen with contrast revealed solid mass and encapsulated cystic precisely identified in the right subhepatic area which assumably formed from retroperitoneum. The lesion is compressing on the following: right lobe of the liver, right branch of portal vein, and hepatic artery, nonetheless, they appear visible.

In addition, the lesion dislodges the main portal vein, superior mesenteric vessels medially, and both medial and inferior bowel loops. Measures of the mass reported 7.2 cm for craniocaudal, 7.2 cm for transverse, and 6.4 cm for AP dimensions, thus evidently suggesting the presence of limbs, vertebral column, and anomalous blood supply to the mass (Figure 3). According to the radiological investigation outcomes FIF was diagnosed and then surgical excision procedure was decided for a complete treatment.

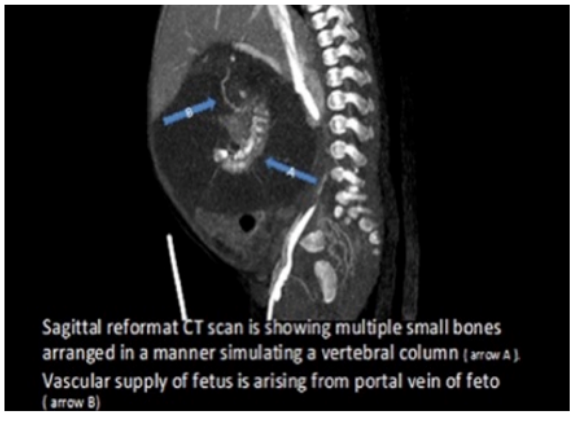

Figure 3: *CT scan, sagittal view.*

Before the surgical procedure the parents were counselled after discussion and signed informed consent was obtained. In the operating theatre, the baby was placed in a supine position with the support of an assistant consultant, and vital signs monitors including: SpO_2 probe, NIBP and electrocardiogram (ECG) were connected. In addition, paediatric endotracheal intubation equipment and laryngeal mask airway (LMA) size 1.0 were prepared and ready to use. Premedication-Inj. Atropine 0.06 mg, Inj. Fentanyl 6 mcg, Inj. Midazolam 0.1 mg and Inj. Ketamine 3.0 mg was injected slowly watching the spontaneous breathing movements.

Initial check with laryngoscopy was done using Miller blade Cormack-Lehane grade 3 view was obtained. Following LMA size number1.0 was easily and successfully inserted, then the patient was ventilated with mixed gases of O_2 and Sevoflurane on JR-circuit along with monitoring SpO_2 and $EtCO_2$. The Anaesthetic plane was deepened with Inj. Propofol and Inj. Cis-Atracurium. The LMA was removed after ventilating for 3 minutes to definite that the airway is secured to facilitate the surgery. In the second stage, the laryngoscope was inserted again for ETT intubation however, the laryngoscopy was difficult. Then Cormack-Lehane grade II was delivered by external laryngeal pressure given by an assistant. Tracheal intubation with Portex ETT size 3.0 mm was confirmed immediately by $EtCO_2$ graph reading. The anesthesia was maintained using mixed gases of N_2O and O_2 along with intravenous Inj. Cis-Atracurium. A Paracetamol suppository of 80 mg was inserted. The surgical area was draped carefully in order to avoid kinking of the ETT. The surgical procedure was uneventful. The trachea was extubated after reversal medication of neuromuscular blockade was given.

Intraoperatively, a surgical incision through right supraumbilical transverse was made, a large retroperitoneal mass was emerged in the subhepatic region. Gallbladder was adherent to the mass and small bowel was pushed toward contralateral side therefore, careful dissection was achieved to release both gallbladder and biliary tree. Furthermore, right hepatic artery which is feeding vessels were controlled uneventfully. Draining veins of the mass into the right portal and superior mesenteric vein were tackled down and ligated successfully, thereafter the mass was completely detached free and removed from the abdomen. Thence, posterior minimal bleeding was controlled with pressure then the wound was sutured in layers. The mass measured 12 x 6 x 6 cm and weighted 690g. In terms of Morphologic, the mass is described as a fetus containing upper and lower limbs with skin covered with hair (Figure 4). For further macroscopic and microscopic investigations, the mass was sent to a histopathology laboratory. Later, the lab results confirmed the

radiological findings of the presence of mature brain tissue, small intestine, and muscle tissue inside the mass. Postoperatively, our patient showed clinical remarkable progress, accordingly was extubated 7 days postoperatively. Moreover, through the period PTH level started gradually dropping down to reach 70 pg/ml. The patient was permitted for discharging after observing sufficient feeding with acceptable weight gain and lowered trend of liver function.

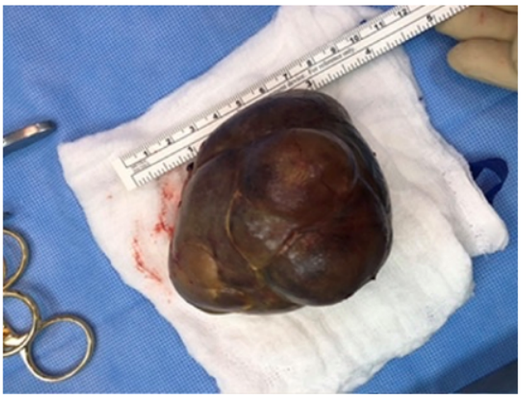

Figure 4: *The mass measures 12 x 6 x 6 cm.*

Discussion

Fetus in feto is a rare malformed condition most reports have diagnosed it through antenatal scan or neonatal period which is characterized by painless abdominal mass [1-4]. Similarly, literature has evidenced reporting this rarity in adults as well [2].

In this case report, an intra-abdominal solid-cystic mass sized 2.6 x 2.6 x 2.3 cm showed in the antenatal scan of the mother. In the postnatal stage, further examinations showed a retroperitoneal mass confirmed the presence of vertebrae and long bone. Yet, FIF condition was reported in different areas of the human body like intracranial [3-5], mediastinum [6] and scrotum [7]. A retroperitoneal Teratoma and FIF were not vividly distinguished until the 1950s Willis established his standard, like the presence of bony parts and spinal column are considered as emphasized on the progressive development and organization of the mass, favoring FIF over Teratoma [8]. However, his standard is universally unknown, and yet it has its drawbacks since it is stressing the presence of axial bony structure and vertebrae as no evidence reported cases containing spinal vertebrae but then proved to be FIF. Kimmel., *et al*. reported a case of a newborn with hydrocephalus disorder diagnosed with intracranial five masses and only two of the five have met Willi"s criteria of the existence of a vertebral column [9]. In this case, skin tissue and hair were covering the mass, and also a section of the mass shows the following of a skull structure with a brain substance, intestines, and bony cartilage (Figure 5).

Figure 5: *Section of the mass shows a skull-like structure with a brain like substance and bony cartilage and intestines.*

In this case that no pericardial or cardiac tissue was found inside the mass, and neither of the literature has reported such findings. Commonly, FIF disorder is believed to be a benign condition, yet reports evidenced some malignant cases were isolated after surgical resection [10]. Accordingly, this has encouraged surgeons for total resection of the mass and then observation postoperatively by tumor markers (Alpha-Feto-Protein and Beta Human Chorionic Gonadotrophin) for a period of two years [10,11]. Thus, surgical excision procedure of the mass is providing a complete treatment in most reported cases. Additionally, our patient of concern was diagnosed with Hyperparathyroidism which explains later diffuse osteopenia and femur fracture (Figure 6). Despite that, postoperatively the x-ray scan showed progression healing of bone fracture and resolution of high PTH, which later clarified that FIF is the source.

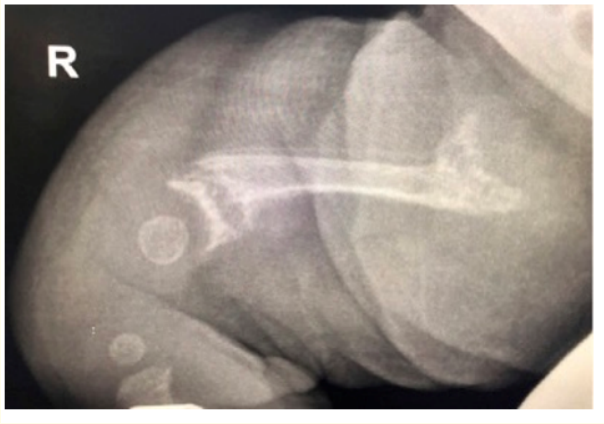

Figure 6: *X-Ray shows right femur fracture.*

Evidence illustrates that the highest incidence rate of conjoint twins is encountered in humans. Conjoined twinning happens when the twinning event occurs at about the primitive streak stage of development, at about 13 - 14 days after fertilization and is associated with the monoamniotic monochorionic type of placentation.

In this case, the preoperative diagnosis was based on clinical and radiological findings in which the presence of cranium, femurs, and vertebrae-like structures was confirmatory [12]. Surgery was critically urgent for main reasons; the difficulty of breathing and feeding and the death of the malformed fetus inside the abdomen. This was considered a privilege compared to other conjoint twins, where sharing of multiple organs is a concern, then accordingly transferring anaesthetic medication through bloodstream would be a core issue. Yet, airway management was more challenging compared to the surgical procedure itself. We have faced two problems, first; a difficult airway caused by the abdominal mass which leads to rapid desaturation, and second; how to maintain ventilation between induction and intubation stages.

Since mask ventilation was difficult, that ruled out the possibility of inhalational induction. Oral Fiberoptic intubation was an option but a convenient size Fiberscope was unavailable, So intravenous induction was only the choice. Ketamine [13] was used for sedation in order to maintain regular spontaneous breathing as it is common to stimulate the respiration process. Since severe micrognathia and dysmorphic features are an issue, in this case, the concept of inserting LMA [14,15] before paralyzing the patient for ETT intubation was only the option. Universally classic LMA is recommended as a safe airway device in many such indications.

Whilst the ventilation [16] was achievable, thus muscle relaxant medication was given, and then LMA had to be removed as fixing it in position was difficult. Hence, the chances of its displacement and kinking during surgical manipulations were high.

Correspondingly, LMA ventilation would be challenging in this case. Furthermore, inserting a throat pack provided patent protection for the trachea and promoted ease with ETT. Check laryngoscopy technique [17] helped us to use of muscle relaxant medication to facilitate tracheal intubation and prevent the possibility of laryngospasm. If a failure in tracheal intubation is expected, LMA could be a solution to proceed with the surgical excision. However, emergency tracheostomy preparation was ready as a part of a contingency and alternative plan.

To sum up, difficult paediatric airway especially for a case like a fetus in fetu can be managed with an appropriate plan.

Conclusion

In the background of the above discussion, a case of FIF with abdominal mass causing difficult airway has been described through this report during prenatal ultrasound examination at 34 weeks of gestation.

We conclude that a difficult paediatric airway like the case of fetus in fetu can be tackled with careful airway assessment, planning with anticipation, proper use of equipment, and safety airway procedure like check laryngoscopy.

Patient Consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Conflict of Interest

The following authors have no financial disclosures (Th A, YS, JA, BK, ER).

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