The management of foregut duplication cyst: Journey from antenatal period of diagnosis to postnatal definitive management along with review of literature.

ABSTRACT

Foregut duplications are rare developmental anomalies and relatively very few cases are diagnosed prenatally on fetal sonography. It is one of the causes of acute respiratory distress in infancy and childhood (1,2). Many theories proposed that foregut duplication occurs due to failure of developing vacuoles to coalesce into the developing esophagus (3,4). The presentation of foregut duplication cysts may be intimately related to esophagus intra-thoracic or completely separated swelling. There may be associated spinal deformities as vertebral defects (5).

Introduction: Maximum cases of intra thoracic foregut duplications occur in posterior mediastinum. Being a rare developmental anomaly with rare anticipation on prenatal sonography, multimodal approach and closed monitoring is very much mandatory. Here presenting a case of such foregut duplication cyst diagnosed on prenatal fetal sonography scan and journey from antenatal presentation to postnatal period till complete surgical repair of lesion done.

Case Report

A 29 weeks of gestation pregnancy referred with fetal scan performed revealing a 18 * 28 * 13 mm simple cystic mass and adjacent echogenic area located in retrocardiac region causing little compression on heart. There was no any other cystic mass noted anywhere. Fetal echocardiography showed normal cardiac activity. Rest of fetal parameters were normal including neurological development and lungs development. There was no evidence of oligohydramnios or polyhydramnios. It was felt that mass was most likely to be a neuroblastoma. Amniocentesis was performed, revealing a normal karyo type and normal amniotic fluid metanephrine levels. It was advised to continue pregnancy and closed follow up was kept with repeated fetal scans to observe the growth of the mass noted. On 32 weeks of gestation there was increase in size of mass noted with more prominent cystic nature. Mass was prominently on right side showing hyper echoic right lung, shifting of mediastinum on left side. There was slight peristalsis suspected in cystic mass , a finding that helped us make a differential diagnosis of foregut duplication. Rest of all fetal scan findings were unremarkable and same as before. Fetal magnetic resonance imaging was done due to unavailability. On further follow up, on 36 weeks of gestation there was grossly increased cystic mass on right side of thorax causing complete compression on right lung, fetal heart shifted to opposite side. On fetal echocardiography there was a structurally normal heart with normal great vessel and venous anatomy with mild physiologic pulmonary insufficiency. There was no pericardial effusion. Mild polyhydramnios was noted. The pregnancy was otherwise uncomplicated. Elective caesarian section was planned with ready back up of all pediatric team and pediatric surgery team. A 2,400 gm baby girl was born at 36 weeks. Baby cried immediately after birth shifted to Neonatal intensive care unit in view of tachypnea and decreased air entry on right side. Hematological evaluation and echocardiography of baby on post delivery was normal. High resolution computed tomography (HRCT) was done immediately on day 1 of life after stabilizing baby physiologically. On HRCT there was huge cystic swelling in thorax on right side with compression of right lung and shifting on mediastinum on opposite side extending in retrocardiac region (Fig.1). On day 2 of life there was dropping of saturation and increased tachypnea considered secondary to compression effects of mass in thorax. On urgent open thoracotomy through 5th intercostal space, there was huge cystic mass with completely compressed right lung (Fig.2). Cyst was occupying almost all hemithorax completely pushing diaphragm inferiorly and separated from diaphragm. The straw colored clear fluid was drained out of the cyst, collapsed cyst showed well demarcation and completely separate cystic swelling closely connected to esophagus without intraluminal connection (Fig.3). Mass could be excised easily without evidence of much adhesions with surrounding tissues achieving complete expansion of lung (Fig.4 & 5). Inter costal drainage tube was kept in situ and thoracotomy wound was closed. Baby got extubated on table with improved physiological and vital conditions. Drain was removed on postoperative day 5. Histopathology study of specimen revealed the multicystic structure features varying layers of circumferential smooth muscle, intervening fibrous tissue containing scattered islands of cartilage and displays a spectrum of foregut derived epithelium and tissues gastric tissues with its mucosa lined by tubular glands which is consistent with foregut duplication cyst (Fig.6). Patient was discharged on post operative day 10 after removal of intercostal drainage tube. On follow up of 4 years of age, baby is achieving normal growth with no any symptomatic presentation.

KEY WORDS: Foregut duplication, Mediastinal mass on prenatal scan, Antenatal management of fetal mediastinal mass.
resonance imaging (MRI) as an adjunct to sonography are of sonography the potential advantages of fetal Magnetic observation are logical. Despite the well-known advantages of differential diagnosis of any intra thoracic cyst. Accurate pathology (16). It should be considered as one of the duplications (15). Intra thoracic gastric duplication is a rare syndrome "postulated the abnormal separation of the notochord by an abnormal orifice and extends to the upper thorax. The prenatal diagnosis of foregut and enteric duplications is very important (12,13). Kassner and colleagues report the case of a young child with an associated pericardial defect abutting an esophageal duplication, though this child's symptoms were aerodigestive rather than cardiac (14).

On prenatal once the diagnosis is confirmed then closed monitoring of pregnancy with serial interval fetal scans are necessary to monitor fetal growth and planning for such pregnancies diagnosed with such lesions. Serial fetal scans are necessary to monitor fetal growth and planning for elective cesarean section depending on physiological stability. Multimodal team approach is always necessary and always be prepared for all emergencies might be during pregnancy or post delivery. Fetal duplications carry a low risk for malignancy, but a definitive diagnosis of a mediastinal mass cannot be determined without histopathology study. Therefore a timely excision of any such lesion after discovery should be considered.

CONCLUSION
Herewith reporting the case which follow of typical pattern associated with proximal foregut duplication but it is highly unique for its size and location in early stage of pregnancy. Herewith would like to give stress on closed monitoring of such pregnancies diagnosed with such lesions. Serial fetal scans are necessary to monitor fetal growth and planning for elective cesarean section depending on physiological stability. Multimodal team approach is always necessary and always be prepared for all emergencies might be during pregnancy or post delivery. Fetal duplications carry a low risk for malignancy, but a definitive diagnosis of a mediastinal mass cannot be determined without histopathology study. Therefore a timely excision of any such lesion after discovery should be considered.

Disclosure
An Informed consent has been taken from parents to publish this case report for academic purpose. I declare no potential conflict of interests, real or perceived.

REFERENCES

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