



ORIGINAL RESEARCH PAPER

THE MANAGEMENT OF FOREGUT DUPLICATION CYST : JOURNEY FROM ANTENATAL PERIOD OF DIAGNOSIS TO POSTNATAL DEFINITIVE MANAGEMENT ALONG WITH REVIEW OF LITERATURE.

Paediatrics

KEY WORDS: Foregut duplication , Mediastinal mass on prenatal scan , Antenatal management of fetal mediastinal mass.

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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 duplications are originating from 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 oligohydromnios or poly hydromnios. 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 pulmonary arterial and venous anatomy with mild physiologic pulmonary insufficiency .There was no pericardial effusion. Mild polyhydromnios 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 5 th 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 five. 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.

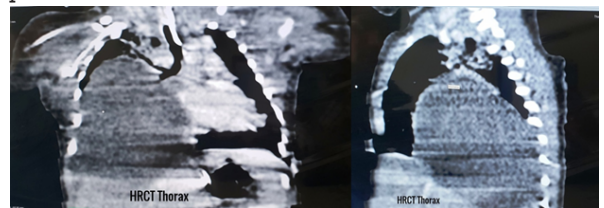


Fig. 1. HRCT Images showing extend of mass and its compression effects on mediastinum.

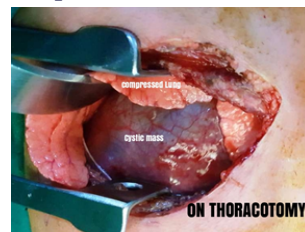


Fig. 2 The mass occupying hemithorax

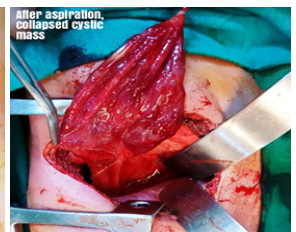


Fig.3 Decompression of Cystic mass



Fig.4. Complete excised specimen



Fig. 5 . Completely expanded lung.

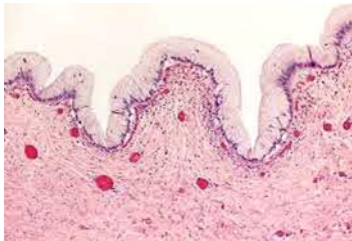


Fig.6 Spectrum of foregut derived epithelium with its mucosa lined by tubular glands on histopathology study.

DISCUSSION

The primary concern in an asymptomatic or minimally symptomatic mediastinal lesion is establishing a diagnosis. Congenital mediastinal masses have malignant potential and operative intervention is advocated.

Majority of mediastinal lesions will be either lymphoma, a neurogenic tumor and germ cell tumor such as teratoma (6,7). Other than these, prenatal suspected mediastinal cystic mass is foregut duplication cyst as differential diagnosis. The risk of malignant transformation in these lesions is very low. Thus far only four cases of cancer arising from foregut duplications have been reported in the literature (8-11). The cyst may present with various sizes, shapes and locations as there may be spherical cyst without communication with bowel lumen, tubular communicating cysts, and sometimes free cystic masses with thin mesenteric stalk. Intestinal duplications are within the bowel mesentery. Foregut duplication cysts are usually in posterior mediastinum in close contact with esophagus and very rarely can be picked up on prenatal scans. Usually esophageal duplications presents in early age up to 6 months of life with respiratory symptoms. On pre natal scan, the another differential diagnosis would include bronchogenic cyst, cystadenomatoid malformation, pulmonary sequestration. Very rare abdomino thoracic variety of duplication cysts may present which communicates with the stomach or upper small intestine, then crosses the diaphragm 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 advocated. Without prenatal diagnosis, an infant may suffer non-specific symptoms like respiratory infections, cardiac problems, failure to thrive etc. for several months before complete evaluation and surgical intervention are undertaken. These infants may experience life-threatening complications if left undiagnosed. As discussed here in case report, as soon as fetal scan presents with complex pregnancy signs then elective caesarian section should be planned. Multi modal approach is always necessary considering requirement of neonatal intensive care support and prompt investigations with surgical intervention.

Even though the etiology of enteric duplication remains unclear, the most widely accepted theory "split notochord syndrome" postulated the abnormal separation of the notochord from the endoderm, leading to enteric duplications (15). Intra thoracic gastric duplication is a rare pathology (16). It should be considered as one of the differential diagnosis of any intra thoracic cyst. Accurate diagnosis is not necessary, but detection and continuous observation are logical. Despite the well-known advantages of sonography the potential advantages of fetal Magnetic resonance imaging (MRI) as an adjunct to sonography are many. Fetal anatomy is well visualized at MR imaging, in part

because of large amounts of amniotic fluid as well as fluid within the fetal lungs, gastrointestinal tract. The multiplanar abilities, large field of view and better tissue characterization of MR imaging may help determine the origin and extent of lesion. MR imaging is not limited by fetal position, fetal bones or maternal body habitus to the same degree as is ultra sonography, particularly in the third trimester (17).

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 scan 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. Foregut 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

1. Gerle RD, Jaretski A, III, Ashley CA, Berne AS. Congenital broncho pulmonary foregut malformation. *N Engl J Med*. 1968;278:1413
2. Rodgers BM, Harman PK, Johnson AM. Bronchopulmonary foregut malformation: The spectrum of anomalies. *Ann Surg*. 1986;203:517-24.
3. Holcomb CW, Gheissari A, O'Neill JA, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. *Ann Surg* 1989;209(2):167-74.
4. Carachi R, Azmy A. Foregut duplications. *Pediatr Surg Int* 2002;18(5e6):371e4
5. Stringer MD, Dinwiddie R, Hall CM, Spitz L. Foregut duplication cysts: a diagnostic challenge. *J R Soc Med* 1993;86(3):174e5.
6. King RM, Telander RL, Smithson WA, Banks PM, Han MT. Primary mediastinal tumors in children. *J Pediatr Surg* 1982;17(5):512e20.
7. Grosfeld JL, Skinner MA, Rescorla FJ, West KW, Scherer 3rd LR. Mediastinal tumors in children: experience with 196 cases. *Ann Surg Oncol* 1994;1(2):121e7.
8. Chuang MT, Barba FA, Kaneko M, Tierstein AS. Adenocarcinoma arising in an intrathoracic duplication cyst of foregut origin: a case report with review of the literature. *Cancer* 1981;47(7):1887e90.
9. Olsen JB, Clemmensen O, Andersen K. Adenocarcinoma arising in a foregut cyst of the mediastinum. *Ann Thorac Surg* 1991;51(3):497e9.
10. Lee MY, Jensen E, Kwak S, Larson RA. Metastatic adenocarcinoma arising in a congenital foregut cyst of the esophagus: a case report with review of the literature. *Am J Clin Oncol* 1998;21(1):64e6.
11. Lecompte JF, Breaud J, Pop D, Venissac N, Mouroux J. Pancreatic adenocarcinoma arising from esophageal duplication. *Ann Thorac Surg* 2012;93(6):2047e8.
12. VanDam, L. J., DeGroot, C. J., Hazelbrock. F.W. and Wladimiroff, J. W. (1984). Intrauterine demonstration of bowel duplication by ultrasound. *Gynecol. Reprod. Biol.*, 18, 229-32
13. Goyert, G. L., Blitz, D., Gibson, P., Seabolt, L., Olszewski, M., Wright, D. J. and Schwartz, D. B. (1991). Prenatal diagnosis of duplication cyst of the pylorus. *Prenat. Diagn.*, 11, 483-6
14. Kassner EG, Rosen Y, Klotz DH. Mediastinal esophageal duplication cyst associated with a partial pericardial defect. *Pediatr Radiol* 1975;4(1):53e6.
15. J. R. Bentley and Smith. J. R., "Developmental posterior enteric remnants and spinal malformations. *Arch Dis Child.*" *Arch Dis Child*, vol. 35, pp. 527-530, 1960
16. P. Daher, L. Karam, and E. Riachy, "Prenatal diagnosis of an intrathoracic gastric duplication: a case report," *Journal of Pediatric Surgery*, vol. 43, no. 7, pp. 1401-1404, 2008.
17. Coakley FV, Glenn OA, Qayyum A, Barkovich AJ, Goldstein R, Filly RA. Fetal MRI: a developing technique for the developing patient. *AJR Am J Roentgenol* 2004;182:243-252.