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RESEARCH ARTICLE

BILATERAL PULMONARY LOBAR AGENESIS – A PROSPECTIVE CADAVERIC STUDY

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ABSTRACT

Bilateral Pulmonary agenesis is a rare congenital anomaly of lungs with a definite embryological basis and diverse Clinical presentations ranging from asymptomatic cases to very severe disease incompatible with extra uterine life. Depending upon the developmental stage of lungs at which the insult occurs, the pulmonary agenesis is classified as bilateral complete, unilateral and lobar agenesis. Diagnosis of this anomaly is possible during early intrauterine life but it may remain silent to be detected at Autopsy. Bilateral lobar agenesis a rare and benign variant of this rare anomaly which we are reporting for the first time in the history of 65 years old Post Graduate department of Anatomy at Government Medical College Srinagar. The present study was carried out during routine dissection for teaching cadaveric Anatomy and simultaneously recording observations.60 lungs from 30formalin preserved Indian cadavers were dissected out and observations regarding the number of lobes in each lung were recorded. In one of the Cadavers we found that right lung had two lobes (upper and lower) and only one fissure (oblique) whereas left lung was single lobed with no fissure. These anomalous lungs were associated with absence of left kidney. No other associated anomalies like tracheal, esophageal, diaphragmatic and cardiovascular anomalies were reported in this case. The sound knowledge of isolated Lobar agenesis is very important in Maternal and child health, Neonatology, Cardiology, chest Medicine, Radio diagnosis and Anesthesiology for early diagnosis and appropriate management of this rare anomaly and its associations which otherwise remain undiagnosed to be detected at autopsy either because of their benign nature or as a consequence of their severity.

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INTRODUCTION

Bilateral complete pulmonary agenesis refers to complete absence of lung parenchyma, blood vessels and branches of tracheobronchial tree beyond the tracheal bifurcation. Depending on its severity it is classified as bilateral complete agenesis, unilateral agenesis and lobar agenesis. The clinical presentation and its severity depend on the appearance time of the malformation during the time line of lung development and the presence or the absence of associated anomalies. This condition must be differentiated from pulmonary hypoplasia in which case the lung parenchyma and pulmonary vessels are under developed with rudimentary principal bronchus. The first reported case of pulmonary agenesis was described by De Pozze in 1673 after autopsy of a woman (Maltz and Nadas, 1968).

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Since then many cases were published yet bilateral and unilateral pulmonary agenesis remains a rare congenital anomaly and unilateral agenesis predominantly affects the left lung in 70% of cases (Dobremez *et al*., 2005) The reported incidence of unilateral pulmonary agenesis is 1 in 15000 live births (Maltz and Nadas, 1968) The exact etiology Pulmonary agenesis is not known. It is usually associated with other congenital malformations in 60 percent of cases (Maltz and Nadas, 1968) and affects both sexes equally (Ootaki *et al*., 2001). Boyden (1955) classified the congenital pulmonary involvement into three groups as pulmonary agenesis, pulmonary aplasia and pulmonary hypoplasia (Kurkcuoglu *et al*., 2005).Unilateral pulmonary agenesis is less severe form of this developmental defect compatible with long term survival. The prognosis of this less severe form of defect depends on the presence of other associated anomalies like cardiovascular (Currarino and Williams, 1985; Finci *et al*., 1999), gastrointestinal (Yancey and Richards, 1993; Steadland *et al*., 1995), and genitourinary malformations (Cunningham and Mann, 1997). Other reported associations of unilateral pulmonary agenesis are ipsilateral skeletal muscle anomalies

(Osborne *et al*., 1989; Aggarwal *et al*., 2002), nonimmunehydropsfetalis and certain chromosomal deletions (Conway *et al*., 2002; Fitoz *et al*., 2001) Bilateral pulmonary lung agenesis is an extremely rare anomaly incompatible with extra uterine life (Vettraino *et al*., 2003).

MATERIALS AND METHODS

The present study was carried out in the postgraduate department of Anatomy Government Medical College Srinagar, Kashmir, India by dissecting30 formalin preserved Indian cadavers for routine teaching purposes and simultaneously recording observations. Dissection of thorax was meticulously carried out strictly following instructions given in the Cunningham's manual of practical Anatomy and observations regarding the number of lobes of 60 lungs were recorded. It was also observed whether anomalies in the number of lung lobes is associated with anomalies of cardiovascular system, tracheobronchial system, urogenital system and diaphragm. Appropriate photographs were taken and labeledas shown in Figs, (1-2).

Fig, 1. Right Lung with only two lobes and absent transverse fissure

RESULTS

After dissecting 30 cadavers and studying 60 lungs, in one middle aged female cadaver we found that the both lungs were malformed. As shown in Figure (Maltz and Nadas, 1968), the right lung had two lobes upper and lower separated by the oblique fissure. The middle lobe and the horizontal fissure were absent. In the malformed left lung no fissure was present making it single lobed. There were no associated anomalies of trachea, diaphragm and cardiovascular system.

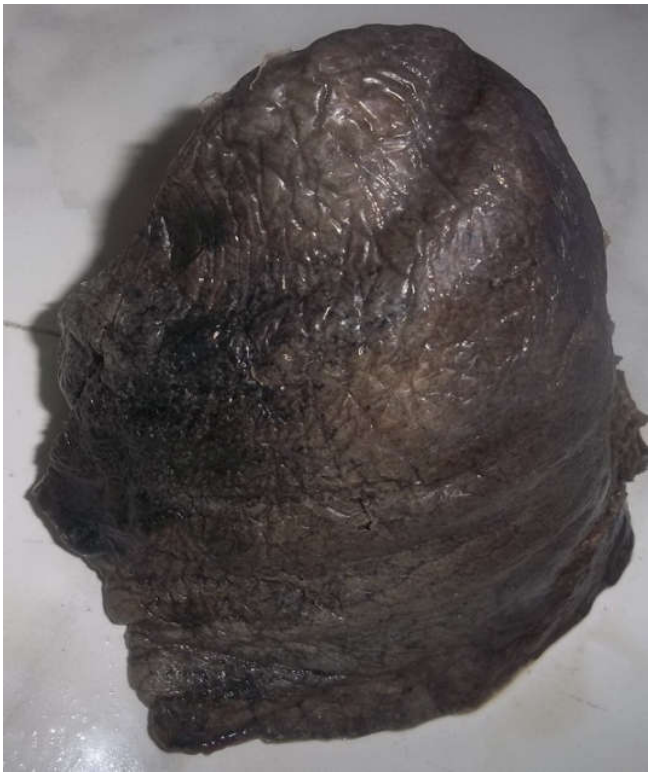

Fig. 2. Costal surface of single Lobed Left lung

This anomaly was associated with agenesis of left kidney and third root of median nerve in the left arm.

DISCUSSION

The respiratory primordium appears around 28 days as a median outgrowth –the laryngotracheal grove - from the caudal end of the ventral wall of the primordial pharynx. From the lining endoderm of laryngotracheal tracheal grove originate pulmonary epithelium and glands of larynx, trachea and bronchi. The connective tissue, cartilage, smooth muscle in these structures develops from surrounding splanchnic mesoderm. The development and maturation of pulmonary tissues follows a definite time table. By the end of the 4th week, the laryngotracheal grooveevaginates to form a pouch like laryngotracheal diverticulum. With progressive elongation of this diverticulum, it is invested by splancnicmesonchyme and its distal end enlarges and forms a globular respiratory bud which gives rise to trachiobronchial tree. During fourth week of intrauterine life the lung bud divides into two out punchings-the primary bronchial buds. These respiratory buds grow laterally into the pericardioperitoneal canals-the future pleural cavaties. Soon they divide into secondary and tertiary bronchi. With further divisions, by interacting with the surrounding splancnicmesenchyme, the bronchial buds differentiate into the bronchi and their ramifications into the lungs. The luminal connection between main bronchus and trachea is established in 5th week. The main bronchus divides and subdivides repeatedly till secondary (lobar) and Segmental (tertiary) bronchi are formed. On the right side, the superior lobar bronchus supplies the superior (upper) lobe of lung. The right inferior bronchus sub divides into two bronchi, one for middle lobe and the other for inferior lobe. In the left lung, the two

secondary bronchi supply upper and lower lobes. The segmental bronchi begin to form by the 7th week. Each segmental bronchus with its surrounding mesenchyme forms primodium of bronchopulmonary segment. Respiratory bronchioles develop by 24 weeks and at this gestational age approximately 17 orders of branches are formed. After birth additional seven orders of trachiobronchial tree develop. During maturation lung passes through pseudoglandular stage (6-16 Weeks), Canalicular stage (16-26Weeks) terminal sac, stage (26 weeks to birth) and alveolar stage (32 weeks to 8 years of age). The first two stages are incompatible with extrauterine life as mechanisms for exchange of gases is not well developed at this stage.

Developmentally a hypoplastic lung results from decreased thoracic capacity as occurs in congenital diaphragmatic hernias which compress the developing lung or as a result of decreased fluid inside developing lungs associated with conditions like oligohydroamnios or renal agenesis. Lung hypoplasia is characterized by markedly reduced lung volume and hypertrophy of smooth muscle in pulmonary arteries**.** Pulmonary agenesis results from failure of respiratory bud to develop. Bilateral agenesis is incompatible with extra uterine life. In unilateral pulmonary agenesis the heart and other mediastinal structures are shifted to the affected side, and the normal lung is hyper expanded. Depending upon the stage of development at which lungs deviate from their normal course of development, the anomalies of the lung scan range from total bronchial and parenchymal agenesis to mild lung parenchymal hypoplasia (Borja *et al*., 1970; Mendelsohn *et al*., 1977) (Borja *et al*., 1970). Most of the published cases had associated congenital defects involving gastrointestinal, genitourinary, cardiovascular, musculoskeltal systems. (Knowles *et al*., 1988; Say *et al*., 1979; Costas *et al*., 1977) (Knowles *et al*., 1988). Being very rare with diverse presentations the exact incidence of this anomaly is not known. Among hospital admissions a prevalence of 0.0034 percent was published by Borja *et al* in 1970 (Knowles *et al*., 1988). There is no reported sex prelidiction of this condition (Stenberg *et al*., 1966) (Stein and Stein, 1966). Morphological classification divides pulmonary agenesis into three different types based on the extent to which bronchopulmonary tissue is absent. Spencer (1977) modified the earlier classifications but presently the classification given by Boyden (Schaffer, 1960) is followed who divided pulmonary agenesis into following types three groups.

Group 1: The lung its pulmonary artery and bronchus are absent.

Group 2: Absent lung and pulmonary artery with a rudimentary bronchus coming off from trachea.

Group 3: Hypoplasic lung with pulmonary vessels and a fully formed bronchus.

60 percent of cases with pulmonary agenesis are associated with other congenital anomalies. Reported associated anomalies are more common with right sided pulmonary agenesis thus accounting for its bad prognosis. The exact etiology of these anomalies and their associations is unclear. For normal development of lungs adequate physical space in the foetal chest cavity and movement of amniotic fluid into the developing lungs are essential. So the factors which decrease the volume of chest cavity like diaphragmatic hernias impair the normal lung development.

The factors which decrease the production of amniotic fluid like renal malformations or impair the movement of amniotic fluid into lungs account for their malformations. According to etiology pulmonary hypoplasia is classified as primary and secondary hypoplasia. The primary is of unknown etiology whereas in secondary hypoplasia either foetal or maternal factors are responsible in 60 percent of the cases. Other causes are space occupying lesions in the chest, developmental anomalies of wall and urogenital and muscular disease (Kurkcuoglu *et al*., 2005) of the chest. Lobar agenesis usually affects right upper and middle lobes (Clements, 1999). Agenesis of a single lobe may remain asymptomatic throughout life to be detected by chest radiograph as an incidental finding or it may bedetectedat autopsy. In pulmonary lobar agenesis theposterioanterior chest radiographic findings include decreased lung volume, shift of mediastinal contents on the affected side and hyperinflated lung on the unaffected side. The lateral chest radiographs may show a band like retrosternal opacity of variable width which is due to shortening of anterio posterior diameter of lung and shift of mediastinal contents (Ang *et al*., 1984). Commonest cardiac malformation associated with lung anomalies are Patent ductusarteriosusand patent foramen ovale (Berrocal *et al*., 2003). The observations made in our present study were different from those made by earlier workers (Borja *et al*., 1970; Knowles *et al*., 1988) who reported that pulmonary agenesis is commonly associated with anomalies of gastrointestinal, genitourinary, cardiovascular, musculoskeltal systems. In our study we found that the lobar agenesis of right and left lungs was associated with absent left kidney and presence of third root of median nerve in the left arm which is not reported in the available literature.

Summary and conclusion

Bilateral lobar agenesis is a rare and benign form of pulmonary agenesis. It may remain asymptomatic or present as recurrent respiratory infections inpaediatricage group. Occasionally lobar agenesis may be detected at autopsy after allowing the affected person to live a normal healthy life. Presently antenatal diagnosis of pulmonary agenesis is possible. There is a definite embryological basis for each variant of pulmonary agenesis. Sixty percent cases of pulmonary agenesis are associated with other congenital anomalies. Isolated anomalies of lung have better prognosis but must be thoroughly investigated to rule out other association.

REFERENCES

Aggarwal, A.N., Gupta, D. and Jindal, S.K. 2002. Unilateral pulmonary agenesis with ipsilateralmusculoskeletal anomalies. *J Assoc Physicians,* India, 50:1324–1326

Ang, J.G. and Proto, A.V. 1984. CT demonstration of congenital pulmonary venolobar syndrome. *J Comput Assist Tomography,* 8:753–7 (Medline).

Berrocalm T,, Madrid, C., Novo, S., Gutienez, Arjovilla, A. and Gomez-Leon N. 2003. Congenital anomalies of the tracheobronchial tree, lung and mediastinum: embryology, radiology and pathology. *Radio graphics,* 24:e17 (Medline).

Borja, A.R., Ransdell, H.T. and Villa, S. Congenital developmental arrest of the lung. *Ann thoracic surg.,* 10:317-326,1970.

Clements, B.S. 1999. Congenital malformations of the lungs and airways. In: Taussing LM, Landau LI, editors. Pediatric respiratory medicine. St Louis: Mosby, p. 1106–36.

Conway, K., Gibson, R., Perkins, J, Cunningham, M.L. 2002. Pulmonary agenesis: expansion of the VCF Sphenotype. *Am J Med Genet.,* 113:89–92.

Cunningham, M.L. and Mann, N. 1997. Pulmonary agenesis: a predictorofipsilatermalformations. *Am J Med Genet.,* 70:391–398

Currarino, G. and Williams, B. 1985. Causes of congenital unilateral pulmonary hypoplasia: a study of 33 cases. *Pediatr. Radiol.,* 15:15–24.

Dobremez, E., Fayon, M. and Vergnes, P. 2005. Right pulmonary agenesis associated with remaining bronchus stenosis, an equivalent of postpneumonectomy syndrome. Treatment by insertion of tissue expander in the thoracic cavity. *Pediatr. Surg. Int.,* 21: 121–122.

Finci, V., Beghetti, M., Kalangos, A. and Brunler, M.A. 1999. Unilateraltotal and contralateral partial pulmonary agenesisassociated with total anomalous pulmonary venous drainage. *J Pediatr.,* 134:510–512

Fitoz, S., Ucar, T., Erden, A. and Gunlemez, A. 2001. Di George syndrome associated left lung aplasia. *Br J Radiol.,* 74:764–766.

Knowles, S., Thomas, R.M., Lindenbaum, R.H. and Winter, R.M. 1988. Pulmonary agenesisas a part of the VACTERL sequence *Arch Dis Child,* 63; 723-726.

Kurkcuoglu, I.C., Eroglu, A., Karaoglanoglu, N. and Polat, P. 2005. Pulmonary hypoplasia in a 52-year old woman. Ann. Thorac. *Surg.,* 79: 689–691.

Kurkcuoglu, I.C., Eroglu, A., Karaoglanoglu, N. and Polat, P. 2005. Pulmonary hypoplasia in a 52-year- old woman. *Ann. Thorac. Surg.,* 79: 689–691.

Maltz, D.L. and Nadas, A.S. 1968. Agenesis of the lung, presentation of eight new cases and review of the literature. Pediatrics., 42: 175–188.

Maltz, D.L. and Nadas, A.S. 1968. Agenesis of the lung: presentationof eight new cases and review of the literature. Pediatrics, 42:175–188

Ootaki, Y., Yamaguchi, M., Oshima, Y., Yoshimura, N. and Oka, S. Pulmonary agenesis with interrupted *aortic arch. Ann. Thorac. Surg.,* 2001; 72: 1742–1744.

Osborne, J., Masel, J. and McCredie, J. 1989. A spectrum of skeletalanomalies associated with pulmonary agenesis: possibleneural crest injuries. *Pediatr. Radiol.,* 19:425432.

Schaffer, A.J. 1960. Pulmonary agenesis and hypoplasia. In Schaffer AJ, Avery ME eds. Disease of the new born. Philadelphia: WB Saunders, 124-209.

Steadland, K.M., Langham, M.R., Greene, M.A., Bagwell, C.E., Kays, D.W. and Talbert, J.L. 1995. Unilateral pulmonary agenesis,esophageal atresia, and distaltracheoesophagealfistula. *Ann. Thorac. Surg.,* 59:511–513.

Stein, I. and Stein, H.L. 1966. Angiograph in the diagnosis of agenesis of lung. *Am J Roentgenol RadiatTherNucl Med*., 96:991-1006.

Vettraino, I.M., Tawil, A. and Comstock, C.H. Bilateral pulmonary agenesis: prenatal sono graphic appearance simulates diaphragmatic hernia. *J Ultrasound Med,* 2003; 22:723–726

Yancey, M.K. and Richards, D.S. 1993. Prenatal sono graphic findings associated with unilateral pulmonary agenesis. *Obstet. Gynecol.,* 81:847–849.

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