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Case Report

AGENESIS OF LEFT LUNG AND LEFT KIDNEY WITH RADIAL RAY ANOMALY

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ABSTRACT

We report a case of 25 yr old female who came to our Pulmonary OPD with insidious left sided chest pain and was proved to have Agensis left lung associated and left kidney, a rare association. On further investigations there is absence of left pulmonary artery and hypoplastic left radius with bowing of ulna.

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INTRODUCTION

Pulmonary agenesis is complete absence of growth of lung. Incidence of unilateral lung agenesis is low. Chest radiograph shows only 1 of 10,000 individuals, (Gilbert *et al*., 1976). Unilateral Pulmonary agenesis is compatible with normal existance. It is associated with anomalies of heart (14%), vascular (9%), genitourinary (9%), gastrointestinal (14%), musculoskeletal system (12%) (Smith and Bech 1958). There is absence of Pulmonary Artery if there is no Lung tissue, this condition is true agenesis (Evans *et al*., 2006), which is present in our case. Renal agenesis consists of absence of one or both kidneys with ureter. Unilateral renal agenesis occurs1-2/1000 births. Presentation is more in males and left side. It is also more common in twins than in singletons (Bianchi *et al*., 2010; Maltz *et al*., 1968). Bilateral Renal agenesis patients are stillborn or die within few days of birth. With unilateral kidney, patient is asymptomatic. Incidence of pulmonary hypoplasia with renal agenesis in neonatal five cases (Osborne *et al*., t989), and infant necropsies (Mirapeix *et al*., 1995). One case report in Spain during adulthood was reported (Hulsoff *et al*., 1959). A case of pulmonary agenesis with absent kidney has not been reported till date, as seen in our case.

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Case Report

A 25 yr old female presented with insidious onset of left sided chest pain since 2 months. There was no cough, breathlessness, orthopnea, palpitation, haemoptysis, anorexia and weight loss. Her perinatal history was insignificant and no history of similar complaints in her siblings. On examination she was an average built female, moderately nourished, preferring right lateral decubitus. She had no pallor, icterus, clubbing, engorged neck veins, lymphadenopathy, Central Cyanosis and Oedema. On Inspection of chest, drooping of shoulder seen in left side and scoliosis with convexity to right was noticed. On palpation respiratory movements were diminshed on left side with rib crowding, trachea deviated to left and apex beat in the left 6th intercoastal space in mid axillary line. Expansion of chest was 2 cm and vocal fremitus diminished throughout the left side. On percussion, resonant note in left infraclavicular area, rest of chest had dull percussion note. On auscultation decreased vesicular breath sounds and decreased vocal resonance in left infraclavicular area was heard. Rest of the areas on left side, breath sounds and vocal resonance were absent. On right side, vesicular breath sound was heard, S2 was loud. Other systems were normal. Chest radiograph should homogenous opacity in the lefthemithorax, Obliterating left costophrenic angle with gross shifting of the mediastinum towards left and scoliosis with convexity to the right, E C G and Echo study was normal.

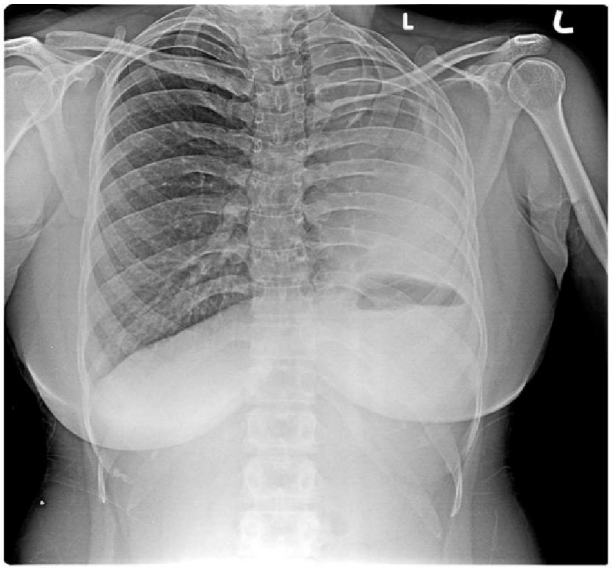

Fig. 1. Chest X-Ray showing homogenous opacity with loss of lung volume in the left hemithorax

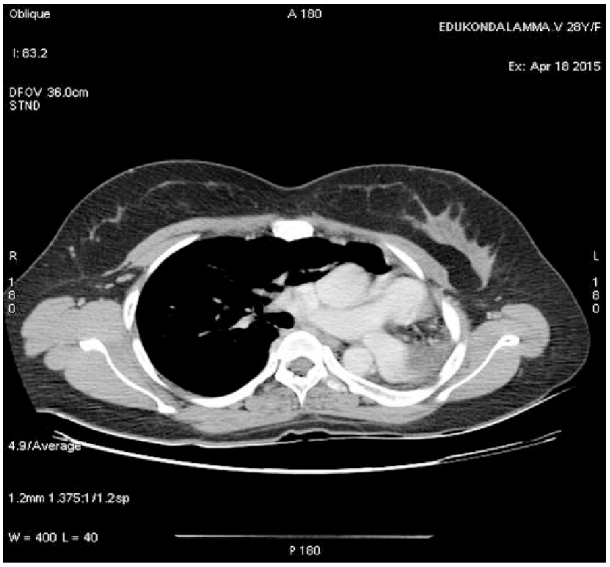

Fig . 2. CECT Chest showing absent left pulmonary artery

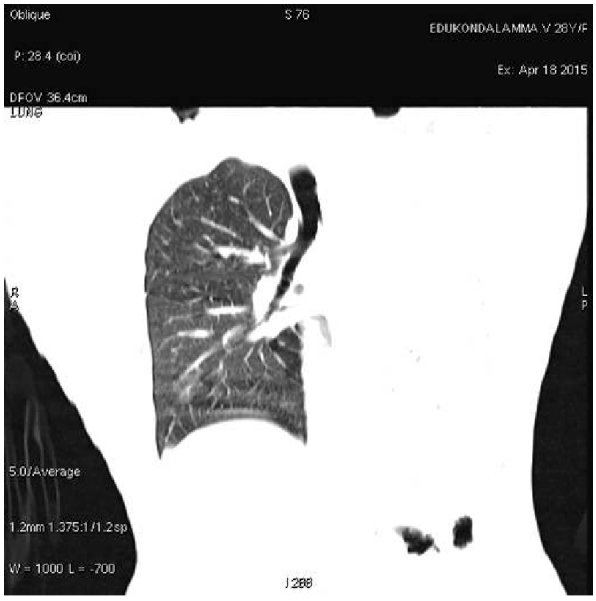

Fig. 3. CT chest suggesting absent left main bronchus

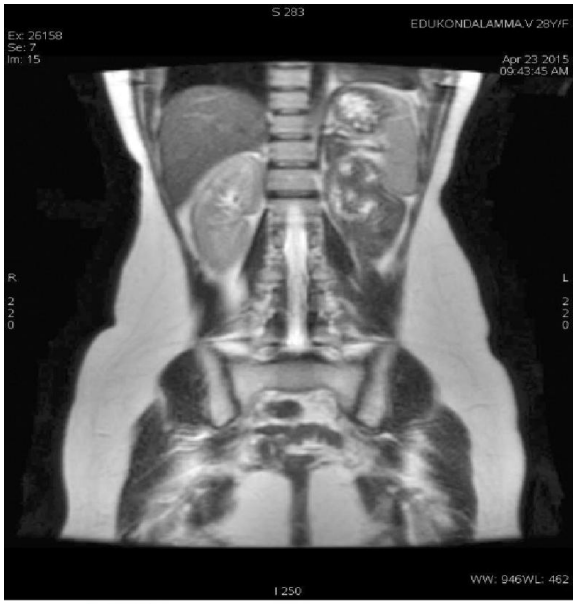

Fig .4. MRI abdomen showing absent of left kidney

Fig.5. Radial ray anomaly

Differential diagnosis in adults with such X-Ray is collapse, thickening of pleura, destroyed lung, pneumonectomy, scoliosis with pleural effusion, diaphragmatic hernia, adenomatoid cystic malformation and Sequestration. To rule these out contrast enhanced computed tomography of chest was done, which showed abscence of left lung and herniation right lung to the left. Left Pulmonary artery was absent. Fibreoptic bronchoscopy showed no opening of left main bronchus. Magnetic resonance imaging of the abdomen confirmed the absence of left Kidney. Xray of left forearm suggested hypoplastic radius with bowing of ulna with 6 carpal bones. We concluded our case to be left lung agenesis with absent left pulmonary artery and left kidney agenesis and radial ray anomaly.

DISCUSSION

Pulmonary agensis has been described in twins and infants with chromosomal abnormalities (Warkany *et al*., 1974; Warkany *et al*., 1995). Patients with unilateral pulmonary agenesis, usually die in neonatal period, survival into adulthood, without symptoms is possible. Arrested development of lung can be classified into three types (Boyden *et al*., 1955).

- Type 1(Agenesis) – Complete absence of lung and bronchus and no vascular supply to the affected side.
- Type 2 (Aplasia) – Rudimentary bronchus with complete absence of pulmonary parenchyma.
- Type 3 (Hypoplasia) – Presence of variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature.

There is no sex predominence. Right and left are equally affected. Right sided agenesis have more cardiac anomalies. Lung development occurs between 3rd and 24th week from median laryngotracheal groove in ventral wall of foregut. By 5th week lung bud starts branching.

Autosomal recessive chromosomal aberration, associated with consanguinous marriage, deficiency of vitamin A, intrauterine infections, environmental factors have been responsible for congenital lung malformation. Renal anomaly is either due to failure of the development of the ureteric bud, or to a defect of its interaction with the metanephric blastema, involving just one side of the urinary tract. The defect in embryogenesis occurs around 5 weeks of embryonic life.

Three factors for renal and pulmonary association

1. Both develop during same period.
2. Both require induction of mesoderm on bronchial bud and on ureteric bud (Larsen *et al*., 1993).
3. Maturation of lung was normal, only in presence of amniotic fluid and intact kidney (Petters *et al*., 1991).

Conclusion

Individual with agenesis of left lung have better prognosis than right. The right lung has excessive displacement of upper and lower medistinum, which hinders drainage of infection and ultimately lung function is affected. With single kidney, infections are recurrent and is prone for calculi formation. This makes regular follow ups necessary (Benjamin Emanuel *et al*., 1974). Presently our patient improved with symptomatic treatment. She has a better life expectancy due to left sided involvement, so we advised her periodic follow ups.

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