Horseshoe lung with left lung hypoplasia

Ali Ersoz, Halim Soncul, Levent Gokgoz, Sedat Kalaycioglu, Sedef Tunaoglu, Melih Kaptanoglu, Ali Yener

Abstract
Horseshoe lung is an uncommon congenital malformation in which the bases of the right and the left lungs are fused to each other by a narrow isthmus posterior to the cardiac apex. So far 22 cases have been described: most of these were associated with right lung hypoplasia and the scimitar syndrome. A horseshoe lung anomaly with left lung hypoplasia is described.

The term horseshoe lung was first used by Spencer in 1962 to describe a malformation in which an isthmus of pulmonary parenchyma extends from the right lung base across the midline behind the pericardium and fuses with the base of the left lung. After this first reported case, 21 more were reported. Most of these cases were associated with hypoplasia of the right lung, dextrarotation of the apex of the heart, and abnormal drainage of the right pulmonary veins into the right atrium.

We describe our experience with a 2½ year old girl who had horseshoe lung with left lung hypoplasia, a ventricular septal defect, and severe pulmonary hypertension.

Case report
A 2½ year old girl was admitted to hospital because of recurrent pulmonary infection, cyanosis, and respiratory failure. Physical examination showed tachycardia, dyspnoea, and central cyanosis. Breath sounds over the left hemithorax were absent. There was a pansystolic heart murmur, mainly at the left sternal border and extending to the apex of the heart. Examination of the other systems showed no abnormalities. On her chest radiograph the left hemithorax was completely occupied by the heart. In the left retrocardiac space atelectatic pulmonary tissue 1.5 cm in diameter was observed. After the surrounding tissues had been dissected abnormally lobulated lung tissue was identified. This could be pulled into the left hemithorax through the inferoposterior cardiac space. There were no hilar structures that belonged to this lung tissue with its abnormal origin. Because full expansion of this atelectatic lung tissue was maintained with positive pressure ventilation resection was not carried out. Instead the lung tissue was fixed to the left hemithorax and pericardium. In the early postoperative period the cyanosis disappeared, blood gas values improved, and a chest radiograph showed that mediastinal shift was nearly corrected and atelectatic segments were re-expanded.

Two weeks after the operation angiography and cardiac catheterisation were performed to investigate the heart murmur. During angiography the left pulmonary artery was not observed, but small collaterals arising from the right pulmonary artery, supplying the left lung (fig 3). During angiography a ventricular septal defect and a patent foramen ovale were
also diagnosed. The mean pulmonary artery pressure was raised (53 mm Hg).

These findings suggested that the child had Eisenmenger’s syndrome, so a medical follow up was preferred. On the 25th postoperative day the child deteriorated with respiratory distress and increased cyanosis. Despite intensive attempts at cardiopulmonary resuscitation she died. Necropsy was not performed.

Discussion

Various cardiovascular anomalies, such as an atrial septal defect, ventricular septal defect, patent ductus arteriosus, and hypoplastic left ventricle, may accompany this rare anomaly. The scimitar syndrome, characterised by abnormal drainage of the right pulmonary veins, dextrotransposition of the apex of the heart, and unilateral lung hypoplasia, has been reported in 15% of 22 cases of horseshoe lung deformity.34 All of 14 cases in a recent series had right lung hypoplasia and 11 of these were associated with the scimitar syndrome.3 Only two horseshoe lung anomalies with left lung hypoplasia had been reported before our case. One of them was diagnosed at necropsy in a 20 week old fetus3 and the other one was in a 2½ year old girl without the scimitar syndrome.3

A possible embryological explanation for this rare anomaly is failure of the caudal splanchnic mesoderm to remain separate and form bilateral organs during the third week of gestation.16

There are no pathognomonic features on the plain chest film in the horseshoe anomaly. The diagnosis can be made by pulmonary arteriography (performed in the lateral and frontal projections), bronchography, or computed tomography, or a combination of these. The appearance of congenital unilateral lung hypoplasia on a chest radiograph and a computed tomogram should suggest the diagnosis. Pulmonary arteriography, which shows the absence of one of the pulmonary arteries with no perfusion of the hypoplastic lung and isthmus, is one of the most valuable diagnostic methods.37

Appropriate surgical management of the anomaly is generally aimed at the correction of the accompanying cardiac anomalies. In the presence of recurrent infection or progressive pulmonary hypertension the resection of the hypoplastic lung and isthmus can be carried out.3

NOTICES

Postgraduate course on surgery of the airways

A postgraduate course on surgery of the airways will be held on 20 and 21 July 1992 at the Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts. Information and application forms may be obtained from the Thoracic Surgical Unit, Massachusetts General Hospital, Boston, Mass 02114, USA (tel (617) 726-2806, fax (617) 726-7667).

Health professions in 1992: the European challenge

A conference entitled “Health Professions in 1992: the European challenge” will be held by the Royal Society of Health on 28 April 1992 at the Guildhall, London (fee £75, reduced for members of the Royal Society of Health). Details from the Conference Department, Royal Society of Health, 38A St George’s Drive, London SW1V 4BH (tel 071 630 0121, fax 071 976 6847).

AIDS in the 1990s

A lecture entitled “AIDS in the 1990s: a global analysis” will be given by Professor Jonathan Mann, Director of the International AIDS Center, Harvard AIDS Institute, on 28 April 1992 at the Guildhall, London. Admission is free but by ticket only, and this may be obtained from the Conference Department, Royal Society of Health, RSH House, 38A St George’s Drive, London SW1V 4BH (telephone 071 630 0121, fax 071 976 6874).