

## CASE FOR DIAGNOSIS

# Repeated lung infections

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### Case history

A 25 year old, nonsmoking white male was admitted to the Emergency Department following suicidal aspirin poisoning. After gastric lavage and X-ray examination, he was transferred to the Pulmonology Department with suspicion of right lung partial atelectasis, probably due to aspiration of a foreign body. He had a history of repeated bronchial infections with dyspnoea, since early childhood. There was no available medical documentation from a period of hospitalization at the age of 12 yrs.

On admission to the Pulmonology Department, the patient was in good general condition. He complained of cough and dyspnoea on exercise. His body temperature was 37.6° C.

Physical examination revealed a narrow right hemithorax, and wheezes and rhonchi, especially in the right

hemithorax. There was a diminished pulmonary function, especially airflow limitation and restrictive changes; total lung capacity (TLC) 79% predicted (pred), forced vital capacity (FVC) 76% pred, forced expiratory volume in one second (FEV<sub>1</sub>) 66% pred, peak expiratory flow (PEF) 58% pred, forced mid-expiratory flow (FEF<sub>50</sub>) 46% pred, carbon monoxide transfer coefficient (K<sub>CO</sub>) 81% pred, static compliance (C<sub>stat</sub>) 66% pred).

A chest X-ray was performed (fig. 1). Perfusion scan showed significantly reduced perfusion to the right lung. During bronchoscopy no foreign body or aspiration mass was detected. To assist diagnosis bronchography was performed (fig. 2). Lung scan and pulmonary angiography were suggested.



Fig. 1. - X-ray at the time of admission.



Fig. 2. - Bronchogram of the right lung.

**BEFORE TURNING THE PAGE: - INTERPRET THE CHEST X-RAY AND THE BRONCHOGRAM. EXPLAIN THE CURVED SHADOW NEAR THE RIGHT HEART BORDER.**

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### Interpretation of the chest X-ray

There was loss of volume of the right lung, with narrowed intercostal spaces and with poorly visualized, diminished right hilar shadow. The trachea and heart are displaced to the right. The right hemidiaphragm was elevated. There was hyperlucency of the left lung, due to compensatory hyperinflation. A poorly visualized curved shadow, near the right heart border, descending to the right diaphragm was also observed (arrows in figure 1).

Computed tomographic (CT) scan confirmed the presence of a small right hemithorax, with reduced vascular markings of the right lung and small right hilum. Herniation of the left lung to the right hemithorax anterior to the mediastinum was also seen.

The bronchogram revealed a narrow main bronchial passage in the middle and right lower lobes, without peripheral small branches. No right upper lobe passage was seen (fig. 2).

Fibreoptic bronchoscopy confirmed the bronchographic findings. No right upper lobe orifice was seen, and main bronchial passage in the medium and lower lobes was narrow and divided dichotomiously. No abnormalities were found in the left bronchial tree.

Pulmonary angiography revealed a small hypoplastic right pulmonary artery, without upper lobe branch. During the venous phase of angiography a poorly contrasted curved shadow was visualized within the heart silhouette. *This shadow was caused by the right pulmonary vein, with entrance into the inferior vena cava near the right leaf of the diaphragm (arrows in figure 3).*

Doppler echocardiography revealed no abnormalities in heart valves and no pathological cardiac shunts.



Fig. 3. — Pulmonary angiograph (venous phase), showing a poorly contrasted curved shadow of the right pulmonary vein with exit into the inferior vena cava, just near the right leaf of the diaphragm (arrows).

### DIAGNOSIS: "Scimitar syndrome"

After oral antibiotic therapy (doxycycline), and inhalation therapy with beclomethasone and fenoterol, the patient was discharged, with clinical improvement.

### Discussion

"Scimitar syndrome" or "hypogenetic lung syndrome" or "Halasz syndrome" [1] is a rare congenital anomaly. The eponym "scimitar syndrome" is a term which has principally been used to describe the appearance of one specific type of anomalous connection of the right pulmonary vein to the systemic venous system, at a point near the level of the right leaf of the diaphragm. The term has been derived from the scimitar-shaped configuration occasionally taken by the right pulmonary vein in its course through the right hemithorax [2]. In some cases, the anomalous vein can be visible roentgenologically, as a broad, gently curved shadow descending to the diaphragm just to the right of the heart.

The classical scimitar syndrome consists of an anomalous right pulmonary vein entering into the systemic venous system below the diaphragm, an anomalous arterial supply to the right lower lung area from a vessel arising from the descending aorta, and an underdevelopment (hypoplasia) of the right lung and right pulmonary artery, with dextroposition of the heart and anomalies of the right bronchial tree [3, 4].

Scimitar syndrome may remain clinically silent for a long time. Some patients have reported repeated haemoptysis or bronchopulmonary infections [4, 5].

The present patient had suffered from repeated bronchial infections, with bronchial obstruction, since childhood. Precise evaluation of X-ray (narrow right hemithorax, approximation of the ribs), and also bronchoscopy and bronchography, excluded aspiration of a foreign body as a cause of the reduced lung volume. Pulmonary angiography, bronchography and CT scan confirmed the hypoplasia of the right lung with anomalous vein drainage. In the hypoplastic lung, we found no cystic malformation, which may be present in some patients [4]. No additional cardiovascular abnormalities (intracardiac shunts) or pulmonary hypertension, as often reported by others [6], were found in our patient.

In differential diagnosis, the acquired condition, Macleod's syndrome, should be taken into consideration. However, in patients with Macleod's syndrome at full inspiration, ipsilateral air-trapping due to bronchial obstruction is observed by roentgenography [4, 7]. We did not observe this sign in our patient. Also, reduction in both ventilation and perfusion are found in perfusion-ventilation lung scans of Macleod's patients.

So-called "mini-hemithorax" - indicated by approximation of the ribs and elevation of the diaphragm - can show different configuration of the rib cage in congenital and acquired conditions. Although the first five ribs on the affected side can be crowded in both conditions, the lower anterior ribs tend to be bilaterally symmetrical with congenital anomalies (as in our patient) but crowded and displaced downwards in patients with acquired disease [4]. In some cases, however, it may be very difficult, even in histological examination, to state with certainty whether the lesion is congenital or the result of early acquired infection [8].

Most patients with scimitar syndrome can be managed conservatively [3, 4]. In some cases, surgical correction of the abnormality, by anastomosis of the anomalous vein to the left atrium, could be considered [3, 9, 10].

**Keywords:** Hypogenetic lung syndrome, lung hypoplasia, scimitar syndrome

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