Rare disease

Swyer–James–MacLeod syndrome with ipsilateral herniation of hyperinflated hyperlucent lung

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Summary

Swyer–James–MacLeod syndrome is characterised by unilateral hyperlucency on chest radiograph with small or normal-sized lung on the affected side and compensatory hyperinflation of opposite lung. Hyperinflation of the affected lung is a very rarely reported entity. An adult female patient, who presented with exertional breathlessness and diagnosed to have hypoplastic left pulmonary artery with hyperlucent, hyperinflated and herniated left lung is described.

BACKGROUND

Unilateral pulmonary emphysema as a clinicoradiological entity was described by Swyer and James in 1953¹ and by MacLeod in 1954.² The reported incidence of this rare disease is 0.01% as per 17 450 surveyed chest radiographs.³ It is characterised by a small or a normal-size of the affected lung and the absence of evidence of bronchial obstruction.² According to literature, abnormally large hyperlucent lung is a rare variant of this rare entity.^{4–6} Here we report a hitherto, unreported rare variant with hyperinflation and retrosternal herniated ipsilateral hyperlucent lung.

CASE PRESENTATION

A 22-year-old girl presented to our department with complaints of recurrent respiratory infections, since childhood

and exertional breathlessness of 6 months duration. On examination, her vital signs and general examination findings were within normal limits. Respiratory examination revealed decreased respiratory movements with diminished breath sounds on the left side of chest with features of shift of mediastinum towards right. Cardiac, neurological and abdominal examinations were unremarkable.

INVESTIGATIONS

She had normal haematological and biochemical parameters. Admission chest radiograph showed hyperlucent left lung with shift of mediastinum to the right, with air-trapping and further rightward mediastinal shift in expiratory radiograph (figure 1A,B). CT of chest revealed small left pulmonary artery with hyperinflated left lung

Figure 1 Inspiratory radiograph (A) showing hyperlucent left lung with shift of mediastinum to the contralateral side and expiratory chest radiograph (B) showing air-trapping and further rightward mediastinal shift.

Figure 2 CT of chest showing (A) hyperlucent (blue thin arrow) and retrosternal herniated left lung (thick green arrow) with air-trapping (thin white arrow) at same level on inspiratory CT (B).

(figure 2A), with air-trapping on expiratory CT (figure 2B) and the striking finding of retrosternal herniation of left lung towards right (figure 2A). CT-pulmonary angiogram showed hypoplasia of left pulmonary artery and vein (figure 3A,B). A diagnostic bronchoscopy revealed no endobronchial obstruction. Echocardiogram showed no cardiac structural anomalies. Ventilation-perfusion scan was done, which showed matched defect in ventilation and perfusion in whole of left lung (figure 4).

DIFFERENTIAL DIAGNOSIS

The differential diagnoses of unilateral hyperlucent lung are given below⁷:

Normal
Increased density of contralateral lung, for example pleural effusion/thickening, consolidation
Technical
Rotation, scoliosis
Soft tissue
Mastectomy
Congenital absence of pectoralis muscle
Poliomyelitis
Emphysema
Compensatory: lobar collapse, lobectomy
Obstructive: foreign body, tumour, MacLeod's syndrome, congenital lobar emphysema
Bullous
Vascular
Absent/hypoplastic pulmonary artery
Obstructed pulmonary artery, for example, by tumour, embolus
MacLeod's syndrome
Pneumothorax

TREATMENT

The patient was diagnosed as a case of variant of Swyer-James-MacLeod syndrome and was managed conservatively.

OUTCOME AND FOLLOW-UP

She and her family were given detailed education regarding the nature and course of the disease and treatment options, and was discharged with advice to be under regular follow-up.

DISCUSSION

This is an uncommon disease with the characteristic radiological feature of unilateral hyperlucency due to air-trapping and loss of pulmonary vasculature. The main mechanism underlying pathogenesis is obliterative bronchiolitis. Reflex pulmonary vasoconstriction and hypoperfusion leading to hypoplasia of pulmonary vasculature also contributes to hyperlucency of the pathologic lung.⁸ These are protective mechanisms to reduce ventilation-perfusion mismatch. This disease presents with variable clinical features. Hyperlucent lung may be an incidental finding in an asymptomatic individual on chest radiograph taken for other indications. Most patients are usually asymptomatic. Cough, haemoptysis, dyspnoea and recurrent respiratory infections can be the alternate presentations. The clinico-radiological features of our patient are similar to those described in earlier reports, except that she had hyperinflation of ipsilateral lung. This pattern of ipsilateral emphysema as seen in our case, is not generally recognised as a radiographic presentation, but such findings have been previously reported as well.⁴⁻⁶ Like our previous authors, we are unable to offer an explanation for this unusual phenomenon and the matter is left open to the scientific community. Ipsilateral herniation of hyperinflated hyperlucent lung as in our patient has never been reported in the literature. The first investigation which is usually done includes inspiratory and expiratory chest radiographs, which show radiolucency of affected lung – the pathognomonic feature of this entity. There can be diminished lung volume on involved side and shift of mediastinum to opposite side during expiration. High-resolution CT is the investigation of choice for confirming the features. It clearly delineates lung parenchyma, pulmonary vasculature and helps to rule out endobronchial obstruction. Prevention and treatment of intercurrent respiratory infection is the main aim of management. Radical procedures like lobectomy and pneumonectomy are usually not done, except when there is recurrent respiratory infection due to diseased lung segment.³⁻⁹ If there is no bronchiectatic component, the prognosis is usually favourable.⁵ Regular follow-up is the cornerstone in the management protocol. The case in our report ran an uneventful clinical course. She was managed conservatively and is under our follow-up for the last

Figure 3 CT pulmonary angiogram sagittal (A) and coronal view (B) showing hypoplasia of left pulmonary artery and vein with diminished peripheral vasculature.

Figure 4 Ventilation-perfusion scan demonstrating matched defects in whole of left lung.

1 year. Three months back, she underwent a laprotomy under general anaesthesia for ovarian cyst, which turned out to be benign. She had no postoperative complications.

Learning points

- Swyer–James–MacLeod syndrome can present as ipsilateral hyperlucent hyperinflated lung
- Development of newer radiological methods in future will lead to detection of more variants
- Such patients can undergo surgeries under general anaesthesia with minimal complications.

Competing interests None.

Patient consent Obtained.

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