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CASE REPORT

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Scimitar syndrome of atypical, rare drainage of venous vessel to the superior vena cava. A case report

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Summary

Scimitar syndrome is a rare and complex congenital anomaly characterized by partial or complete anomalous pulmonary venous return from the right or left lung into the inferior vena cava, through drainage into the hepatic vein, right atrium or left atrium. The syndrome is commonly associated with hypoplasia of the right lung and right pulmonary artery. We present an 11-year-old female with atypical and rare type of scimitar syndrome. The girl has had cough for 2 months before admission, without fever or abnormalities on medical examination. X-ray films revealed inflammatory and atelectatic changes with mediastinal shift to the right. CT and CT angiography – hypoplasia of the right lung with no visible interlobar fissures. No areas of consolidation in the pulmonary parenchyma. Mediastinum shifted to the right. Single wide venous vessels draining the upper part of the right lung entering the superior vena cava. In our patient, clinical symptoms are mild, but a thorough physical examination could have helped diagnose the syndrome earlier.

Key words: scimitar syndrome • pediatric • CT angiography

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Background

Scimitar syndrome is a rare and complex congenital anomaly. The true incidence is not known, because the syndrome may be undetected in asymptomatic patients who have not undergone chest radiography. It is characterized by partial or complete anomalous pulmonary venous return from the right or left lung into the inferior vena cava, through drainage into the hepatic vein, right atrium or left atrium [1–4]. The anomaly of pulmonary veins most commonly affects the right lung. In two-thirds of cases, the scimitar vein provides drainage for the entire right lung, but in one-third, this vein drains only the lower portion of the right lung [5–7]. The syndrome is commonly associated with hypoplasia of the right lung and right pulmonary artery, systemic arterial blood supply to the right lower lung from the infradiaphragmatic aorta, atrial septal defect (e.g. *ostium secundum*), right-sided diaphragmatic hernia, persistent left superior vena cava, horseshoe lung and dextroposition of the heart [5]. Two main forms of scimitar syndrome have been described. The infantile form generally presents within the first 2 months of life with tachypnea, recurrent pneumonia, failure to thrive and signs of heart failure [3]. Those with infantile type usually have a severe form of the disease and worse prognosis [7,8]. Diagnosis of the adult form is generally made incidentally and some patients may be completely asymptomatic or present with recurrent respiratory infections and exercise intolerance [1,3,6,8]. We present a patient with atypical and rare type of scimitar syndrome with drainage of venous vessel to the superior vena cava.

Case Report

A previously healthy 11-year-old female was admitted to the Central Clinical Hospital of the Ministry of Internal Affairs with persistent right-sided inflammation and atelectasis on X-ray. Two months before admission, the girl developed cough, without fever or abnormalities on medical examination. Chest radiograph performed after a month,

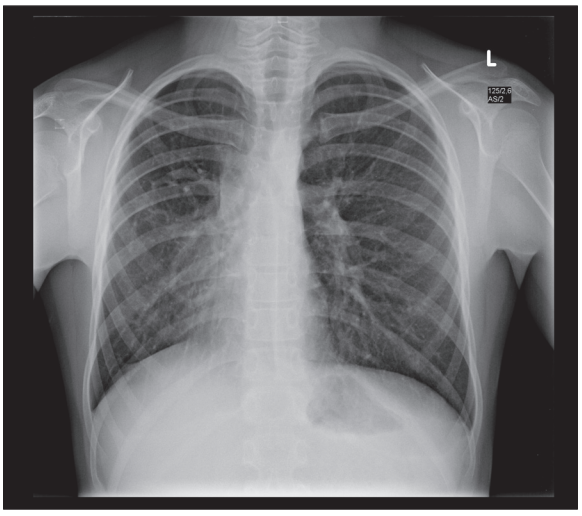

Figure 1. Chest X-ray. Features of right lung volume reduction with mediastinal shift to the right.

showed right-sided inflammation, atelectasis and dextroposition of the heart and trachea. Clarithromycin therapy was prescribed for 24 days. During the treatment, cough relief and regression of pulmonary inflammation were achieved, but atelectasis remained present. There was no evidence of choking, aspiration or contact with coughing or infected persons. On admission, physical examination revealed scoliosis, right-sided cervical lymphadenopathy (approx. 1.5 cm), a shift in the heart sounds and cardiac impulse to the right. There were no other abnormalities. Laboratory tests were normal, serologic tests for *Mycoplasma pneumoniae, Chlamydia pneumoniae, Bordetella pertussis* and *Legionella pneumophila* came back negative. Abdominal ultrasonography and spirometry were normal. Echocardiogram confirmed heart shift to the right and showed anomalous right upper pulmonary vein drainage to the inferior vena cava. X-ray examination revealed inflammatory and atelectatic changes with mediastinal shift to the right (Figure 1). CT angiography – hypoplasia of the right lung with no visible interlobar fissures. No areas of consolidation in the pulmonary parenchyma. Mediastinum shifted to the right. Single wide venous vessels draining the upper part of the right lung entering the superior vena cava (Figure 2). Following consultations with cardiothoracic surgery, pediatric cardiology and pulmonology teams, it was decided that the best course of management would be to follow the patient clinically with serial echocardiography, as there was no significant right-to-left shunt.

Discussion

Scimitar syndrome belongs to a variety of congenital disorders known as "partial anomalous pulmonary venous connections" (PAPVCs), which are found in 0.4–0.7% of the general population [3]. Scimitar syndrome represents about 3–5% of all PAPVCs. It is more common in females and can be familial. [5,7–9]. Etiology of scimitar syndrome is not clear. However, it is considered to be due to an abnormality in the pulmonary development in early pregnancy. Embryonic connection between the pulmonary and systemic veins may persist, resulting in pulmonary venous drainage abnormality [10].

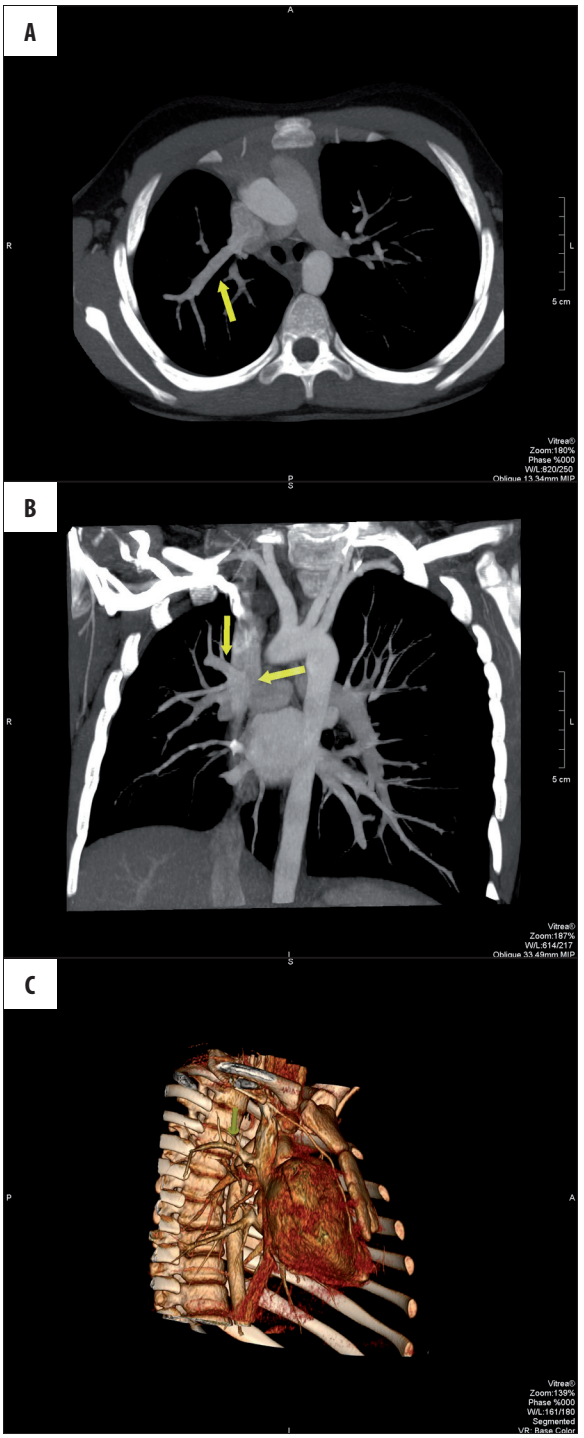

Figure 2. CT angiography. In the reconstruction 2D MIP in axial (**A**), coronal plane (**B**) and 3D VR (**C**) wide anomalous venous vessel (yellow arrows) drains into the superior vena cava, mediastinal shift to the right – dextrocardia.

Variety of symptoms and the presence of any form of the syndrome make it difficult to diagnose. The diagnosis of childhood/adult form is often made incidentally, and indeed, some patients may be completely asymptomatic. Symptoms of patients with Scimitar syndrome depend on

shunt volume and include exercise intolerance, dyspnea, fatigue, palpitations, syncope and congestive heart failure. The most common symptoms during childhood and adolescence are recurrent and prolonged pulmonary infections and dextroposition of the heart as observed in our patient. Other symptoms, such as extreme fatigue, shortness of breath, mild exertional dyspnea, heart murmur and deformation of the right hemithorax, can occur.

Diagnosis of scimitar syndrome is usually made based on characteristic chest X-ray films and can be confirmed by angiography [8]. However, only 70% of these patients have signs of the disease on chest radiography, because this type of Scimitar syndrome does not have right pulmonary hypoplasia [6]. Echocardiography provides a more accurate assessment of abnormal veins and arteries and their relation to the great vessels and the heart. Echocardiogram is also necessary due to the presence of associated cardiac defects in 36% of patients in the pediatric population and up to 76% of neonates. In addition, echocardiography can assess for the presence of pulmonary hypertension, which is very important when deciding on the management. Cardiac catheterization remains the gold standard for diagnosis confirmation and provides a quantitative assessment of the ratio of pulmonary to systemic flow (Qp/Qs). It also provides the opportunity for therapeutic intervention [8]. Due to the lack of features of pulmonary hypertension in echocardiography, we refrained from performing cardiac catheterization. CT, CT angiography and MRI angiography allow for non-invasive diagnosis of Scimitar syndrome and the associated defects. In our patient, we revealed hypoplasia of the right lung with no visible interlobar fissures and single wide venous vessels draining the upper part of the right lung entering the superior vena cava. It is a very rare form of the syndrome [3]. As in our case, patients with no clinical symptoms, no associated defects, with asymptomatic right-to-left shunt and no pulmonary hypertension can be treated pharmacologically [7,8].

In our patient, the clinical symptoms are mild, but a carefully performed physical examination could have helped diagnose the syndrome earlier. Our case illustrates the importance of an accurate and thorough clinical examination of the patient. In the era of highly specialized diagnostic techniques, we cannot forget the basic, old physical examination.

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