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Late Diagnose "Scimitar Syndrome" Associated with Multiple Disorders: Tracheal Stenosis, Tracheomalacia, Chronic Obstructive Pulmonary Disease, Vertebral Malformation

KEYWORDS: scimitar syndrome, multidetector CT, angiography CT imaging, comorbidities

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ABSTRACT *We report the case of a 60 years old male patient (ex-smoker), admitted in our clinic with pneumonia and respiratory failure. CT relevated a mat retracted right hemitorax, left lung hernia, dextro-concave scoliosis with vertebral malformation. Bronchoscopy diagnosed a complex tracheal stenosis, severe tracheomalacia, upper and middle right lobe atresia and a suppuration in the lower right lobe. Echocardiography found a mitral failure. CT angiography confirmed right pulmonary artery atresia and the anomalous connection of the right pulmonary veins into inferior vena cava. We considered a „scimitar syndrome" with late clinical onset and multiples comorbidities. The patients followed treatment for pneumonia, COPD and respiratory failure. As for the tracheal stenosis and tracheomalacia we recommended evaluation in an interventional bronchoscopy service in order to consider tracheal dilations or stent insertion to prevent further complications. Particularities of the case: late, surprising diagnosis of the congenital disease, association with smoking, COPD and other disorders causating respiratory failure (tracheal stenosis, traheomalacia, pneumonia), the decisive role of CT angiography and multidetector CT in elucidating the diagnosis.*

"Scimitar syndrome" was the name assigned for the entity "congenital pulmonary venolobar syndrome" (CPVS) by Neill C.A. et al. in 1960, previous described in 1836 by Cooper G. and Chassinat R. (Cirillo 1998). The name „scimitar" (derived from the persian word for sword) notes the chest X-ray image of the anomalies of the right pulmonary veins draining in the inferior vena cava (curved blade sword like). The "scimitar syndrome" is a rare congenital diseases which includes variable anomalies: hypoplasia of the right pulmonary artery, hypoplasia of the right lung or sequestration, anomalies of bronchial branching, pseudodextrocardia, cardiac malformation in 25% (Godwin&Tarver 1986), anomalous connects between the right pulmonary veins and inferior vena cava with a left-to-right shunt.

The disease is often asymptomatic and compatible with long life, but is associated with dyspneea, fatigue and repeated chest infection (Godwin&Tarver 1986). CT angiography or magnetic resonance imaging can be helpful by depicting the vascular and lung anomalies (Gavazzi, Ravanelli, Farina, Chiari & Maroldi, 2008; Baxter,McFadden, Gradman&Wright 1990; Lee, Boiselle&Cleveland, 2008).

CPVS necessitates surgical intervention in case of severe left-to-right shunt or cardiac malformation. The shunt can be treated by redirecting the scimitar vein into the left atrium. Surgery may be necessary (lung resection) also if there occurs repeated infection in the hypoplasic lung (pneumonia or recurrent supuration). (Brown, Ruzmetov, Minnich & et al 2003; Kamler, Kerkhoff, Budde & Jakob, 2003).

Case report

A 60 years old patient (an ex-smoker male - 20 packages/years) was admitted with respiratory failure. Physical examination disclosed stridor, severe rest dispneea, cyanosis, chest pain, fever, depression of the anterior neck region, reduced basal vesicular murmur, wheezing, sibilantes and crackles, mucopurulent sputum, arrhythmic heart beats.

As a child our patient suffered a tracheostomy and tracheal intubation for a severe episode of respiratory failure supposed induced by a foreign body inhalation and secondary infection. After that cured episode he was asymptomatic long time but in the last two years he had recurrent pneumonia.

The chest-Xray and the chest CT showed a mat retracted right hemithorax with mediastinal shift to the right, left lung hyperinflation with lung haernia, upper tracheal stenosis, traheomegalia, diaphragm raising, dextroconcave scoliosis, vertebral malformation (spinal block). (Figure 1,2,3)

We performed a multidetector chest CT (MDCT) with spatial lung and tracheobronchial reconstruction wich confirmed the chestXray suspicion of tracheomegalia, bronchial and partial right lung atresia and the herniation of the left lung. (Figure 4)

Bronchoscopy diagnosed a complex tracheal stenosis (>2cm length, lumen reduction), followed by a severe tracheomalacia, the absence of the upper and middle right lobe bronchus and a lower right lobe suppuration.

Heart ultrasound found a mitral failure with arrhythmia and a moderate pulmonary hypertension.

Pulmonary function tests showed a severe mixed ventilatory dysfunction.

CT angiography confirmed right pulmonary artery atresia with anomalous connection of the right pulmonary veins into inferior vena cava, right lung hypoplasia and pseudodextrocardia. (Figure 5).

We established the following diagnostic:

1. "Scimitar syndrome" - lung malformation with partial lung atresia and lower right lobe hypoplasia; right pulmonary artery atresia; pseudodextrocardia, mitral failure);

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2. Upper tracheal stenosis posttracheostomia; traheomalacia;
3. Recurrent basal pneumonia; chronic obstructive pulmonary disease (COPD) with exacerbation of chronic respiratory failure;
4. C2-C4 vertebral block; dextroconcave scoliosis;
5. History of chronic smoking.

The patients followed treatment for lung and heart complications (antibiotics, antiarrhythmics), treatment for exacerbated COPD (inhaled bronchodilators, systemic corticoids in short cure, oxygen). At discharge we recomanded bronchodilaters (long acting inhaled anticholinergic, long acting inhaled beta agonist), long-term oxygen if hypoxemia persist, vaccination, pulmonary rehabilitation. The patients had a favorable clinical evolution but he has a poor prognostic, due to the persistence of the severe respiratory failure and the high risk for recurrent infections. As for the tracheal stenosis and tracheomalacia we recommended large evaluation in an interventional pulmonology service in order to establish if tracheal stent insertion is necesary to prevent further complications. The patient refused further investigation.

Discussion

This case has several particularities to discuss and remember:

1. Association of several comorbidities confers a high level of severity:

- Congenital malformation: "scimitar syndrome" and vertebral malformation
- Complex tracheal stenosis post tracheostomia and intubation. The question is if the tracheostomia performed in the childhood for so-called "inhaled foreign body" was not actually an episode of infection with respiratory failure superimposed over the lung disorder;
- Traheomalacia secondary to infections favored by intubation and malformation;
- Severe respiratory failure by different mechanism: restrictive dysfunction by lung atresia and scoliosis and obstructive dysfunction by tracheal stenosis, traheomalacia and COPD;
- Chronic smoking.
- Recurrent respiratory infections favoured by all the comorbidities;

2.Particularities of case evolution

- Late diagnosis of a congenital disorder;
- Long tolerance of severe comorbidities to a smoker.

3. Characteristics of therapeutic approaches

- The complex tracheal stenosis and subsecvent tracheomalacia would recommend the case for tracheal dilations, tracheal stenting or surgical treatment;
- The anesthetic and operative risk are significant due to respiratory and cardiac comorbities;
- Surgery may be necessary (lung resection) if there occurs repeated supuration in the hypoplasic lung). (Brown, Ruzmetov, Minnich & et al 2003; Kamler, Kerkhoff, Budde & Jakob, 2003)

Conclusion

The presented case demonstrates the difficulties that may be encountered in the diagnostic CPVS. CPVS may be asymptomatic as long as complications like infection and respiratory failure do not appear. Early diagnose of the congenital disorder could contribute to an appropriate education and prophylaxis of risk factors for respiratory complication (COPD, infections, smoking): smoking advice, avoidance of the tracheostoma, vaccination, exposure avoidance. Modern CT angiography and multidetector CT are useful in confirmation of associated cardiac, vascular and lung anomalies and there are mandatory in the preoperative approach.

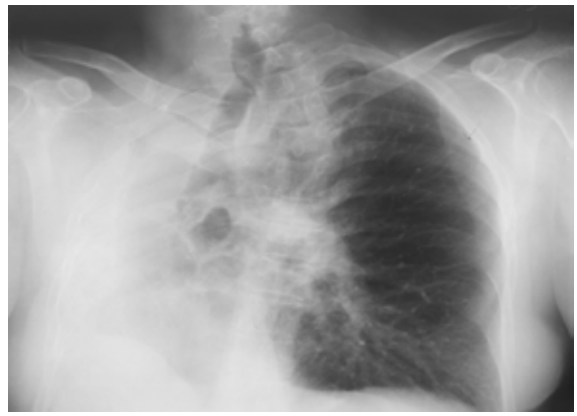

Figure1. Chest X-ray showing a mat retracted right hemithorax with mediastinal shift to the right, left lung hyperinflation with lung haernia, upper tracheal stenosis, tracheomegaly, diaphragm raising and dextroconcave scoliosis

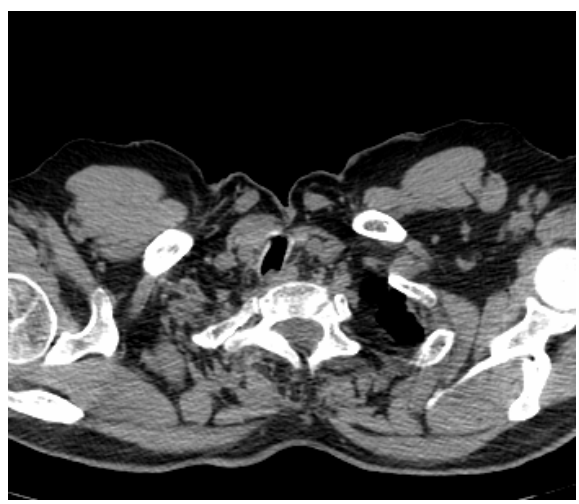

Figure 2. Chest CT, upper sections.

Intrinsic tracheal stenosis and right retraction of the tracheea.

Skin depression coresponding to the cutaneous scar post-tracheostomy

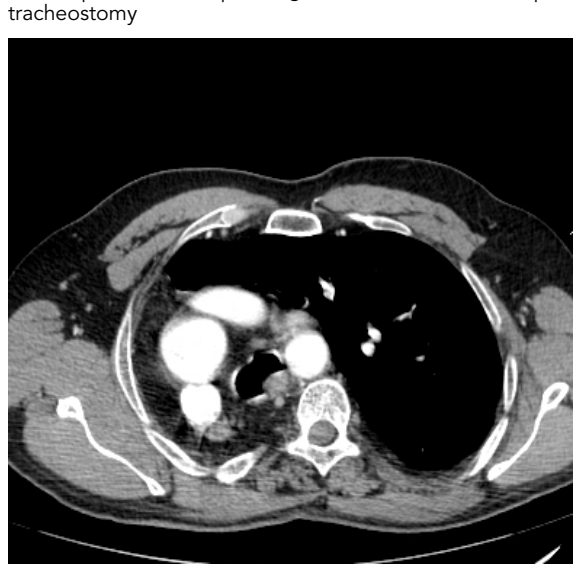

Figure 3. Tracheomegaly. Right cardiac and mediastinal

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shift, right lung atresia, left lung haernia in the right side of the thorax

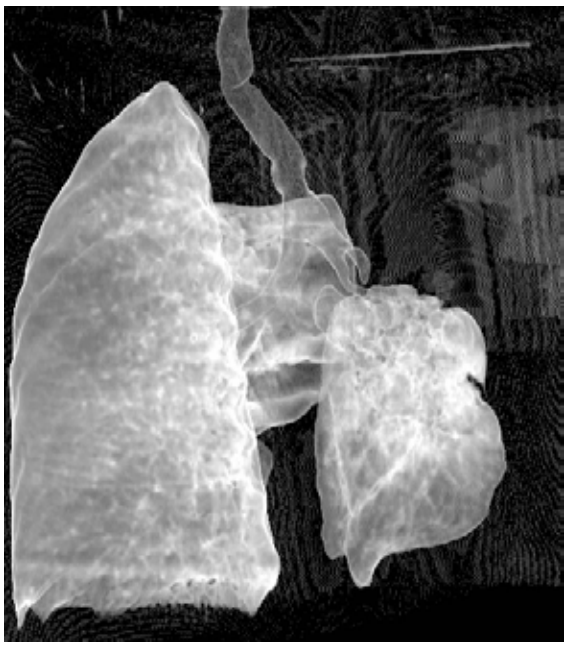

Figure 4. Right lower lobe rudiment with condensation, right and midlle upper lobes atresia, tracheomegaly, tracheal stenosis left lung hyperinflation and herniation to the right

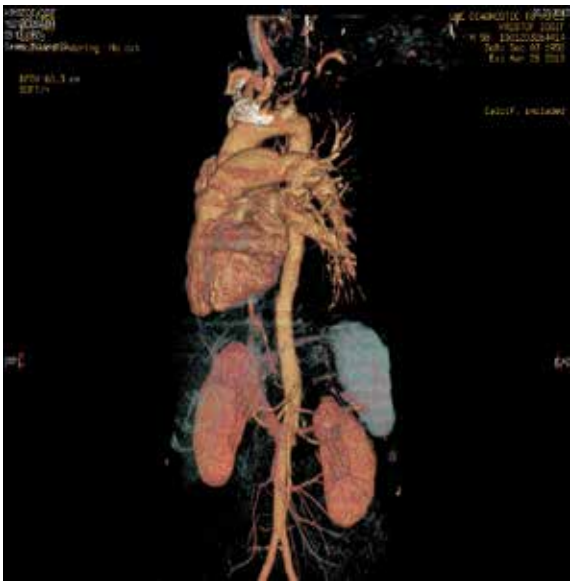

Figure 5. CT angiography. Right pulmonary artery atresia

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