Case 12731

Eurorad

A case of scimitar syndrome in a young woman

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Section: Chest imaging
Area of Interest: Arteries / Aorta Pulmonary vessels Lung
Procedure: Computer Applications-Detection, diagnosis
Procedure: Education
Imaging Technique: CT-Angiography
Imaging Technique: CT
Imaging Technique: Digital radiography
Special Focus: Congenital Case Type: Clinical Cases
Authors: Vinciguerra M, De Dominicis C, Briani C, David V
Patient: 21 years, female

Clinical History:

A 21-year-old woman has been hospitalized for right hydronephrosis and shortness of breath; XR chest showed a diffuse opacity with disappearance of cardiac contours to the right; a chest CT revealed absence of right upper lobe, hypoplasia of right pulmonary artery and two abnormal contiguous vessels.
Imaging Findings:

The chest radiograph showed a diffuse opacity of the right lung with a dextroposition of the heart.
A biphasic (arterial and venous phase) contrast enhanced CT examination of the chest revealed an asymmetry of the chest with a deviation of the mediastinum to the right due to an absence of the right upper lobe. Furthermore, the CT showed an asymmetry of the pulmonary arteries (APD right 11 mm vs APD left 14 mm). At the basal pyramid of the lower lobe two abnormal contiguous vessels were identified, a threadlike vein draining into the inferior vena cava (scimitar vein) and a small artery originating directly from the abdominal aorta corresponding to a systemic arterial supply of the right lower lobe.
In addition, an aberrant subclavian artery was found.
Discussion:

Scimitar syndrome or hypo-genetic pulmonary syndrome is a congenital form of anomalous pulmonary venous drainage (APVD) associated with other anomalies such as hypoplasia of the right lung, dextroposition of the heart, hypoplasia of the right pulmonary artery (RPA) and anomalous systemic arterial supply from the aorta to the right lung.
It was described for the first time by Cooper in 1836 and the term scimitar syndrome was coined by Naill in 1960, because of the radiographic appearance of the anomalous vein (a tubular opacity paralleling the right heart border resembling a curved Turkish sword or Scimitar). [1, 2]
In these patients, pulmonary venous drainage is anomalously connected to systemic vein/s, typically to the superior or inferior vena cava (SVC or IVC) or directly to the right atrium (RA).
In our case, to understand the cause of respiratory symptoms without signs of infection, a chest X-ray followed by a chest CT led to the diagnosis.
Since she was born, our patient was followed for multiple anomalies such as right lower limb deformity, right kidney

anomaly and asymmetry of the pulmonary arteries.
This rare anomaly has an incidence of approximately 1 to 3 per 100, 000 live births. Two different groups of patients exist: adults and older children usually diagnosed during a workup for dyspnoea, fatigue, recurrent respiratory infections or as an incidental finding on a routine chest radiograph, and infants, who become symptomatic soon after birth.
The adult form usually is not associated with pulmonary hypertension and typically has mild symptoms and benign prognosis, the form found in younger patients is often complicated by pulmonary hypertension and cardiac failure. [1, 2]
When necessary, treatment consists of elimination of aortic collaterals by embolization with spirals or surgery by right thoracotomy access.
Differential Diagnosis List: Scimitar syndrome, Pulmonary hypertension, Pulmonary hypoplasia

Final Diagnosis: Scimitar syndrome

References:

M. Kahrom, H.Kahrom (2009) Scimitar syndrome and evolution of managements. Pan Afr Med J 17;3:20 (PMID: 21532729)
Odenthal C, Sarikwal A (2012) Anomalous unilateral single pulmonary vein versus scimitar syndrome: Comparison of two paediatric cases and a review of the literature. J Med Imaging Radiat Oncol 56(3):247-54. doi: 10.1111/j.1754-9485.2012.02385.x (PMID: 22697320)

Figure 1

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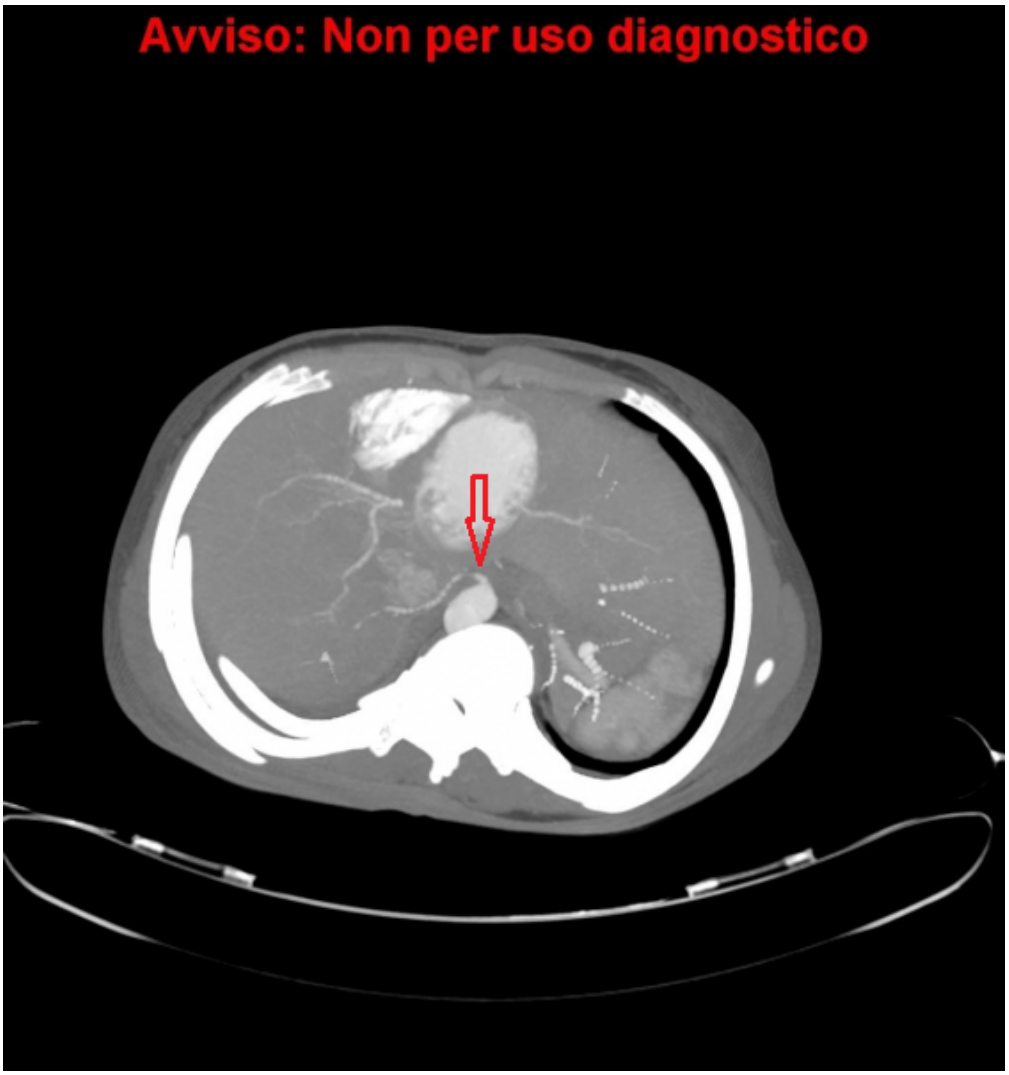
Description: At the basal pyramid of the lower lobe two abnormal contiguous vessels can be identified, a threadlike vein draining into the inferior caval vein and a small artery originating directly from the abdominal aorta. **Origin:** Department of Radiology, Azienda Ospedaliera Sant\'Andrea, University La Sapienza, Rome

Figure 2

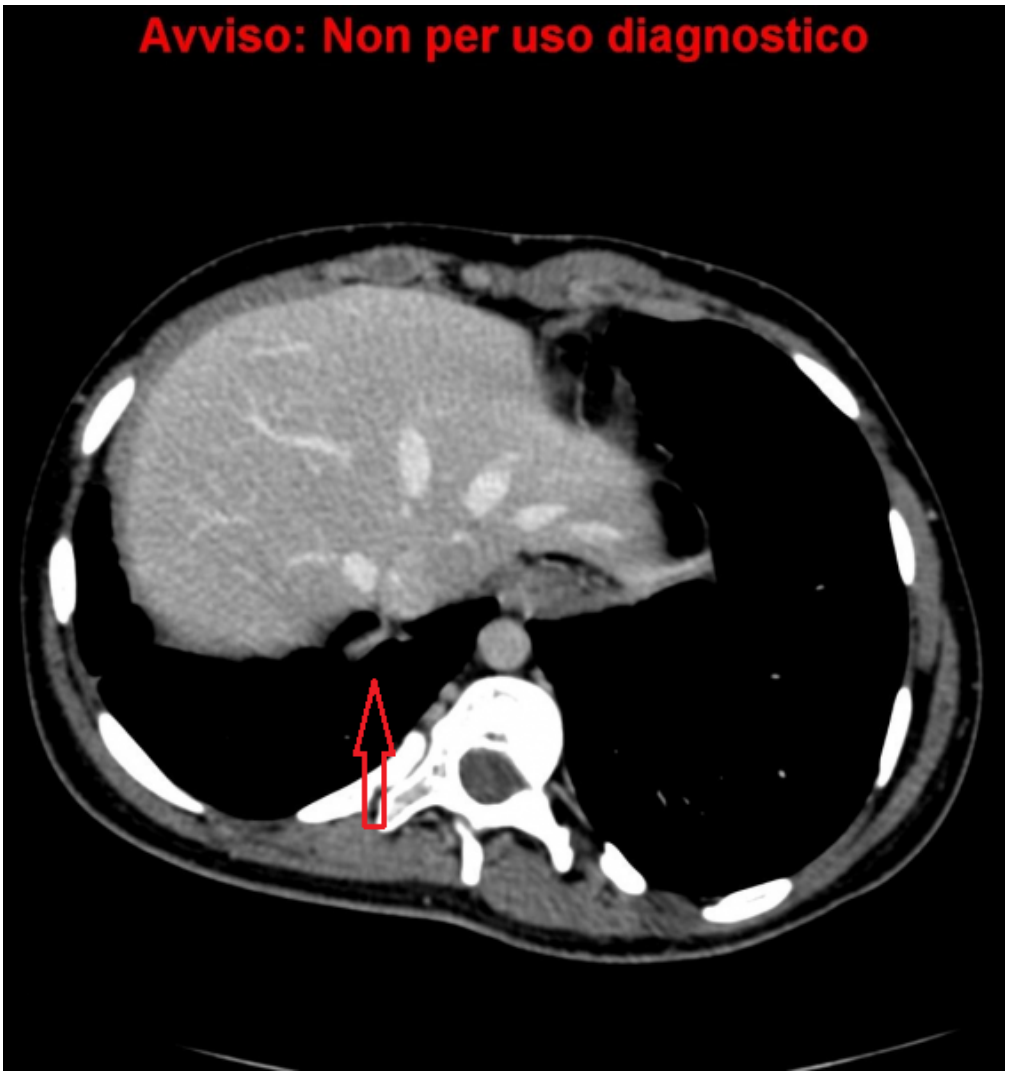
a



Description: Small artery originating directly from the abdominal aorta corresponding to a systemic arterial supply of the right lower lobe. **Origin:** Department of Radiology, Azienda Ospedaliera Sant\'Andrea, University La Sapienza, Rome

Figure 3

a



Description: Vein at the basal pyramid of the right lower lobe draining into the inferior vena cava (scimitar vein). **Origin:** Department of Radiology, Azienda Ospedaliera Sant\'Andrea, University La Sapienza, Rome

Figure 4

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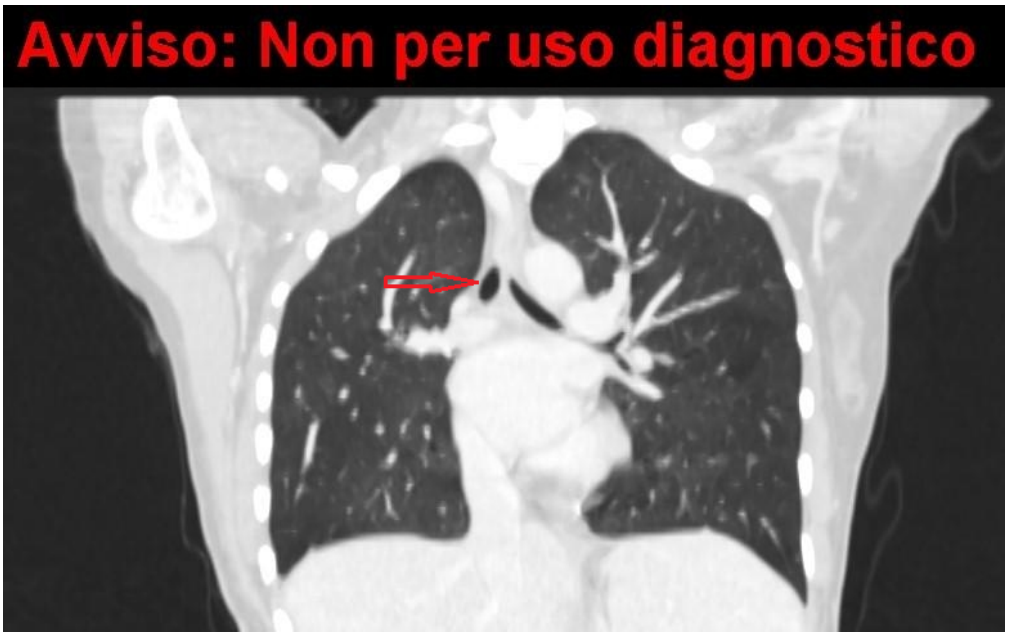
Description: Aplasia of the right upper lobe with consequent loss of volume of the ipsilateral lung and deviation of the mediastinum to the right. **Origin:** Department of Radiology, Azienda Ospedaliera Sant\'Andrea, University La Sapienza, Rome

Figure 5

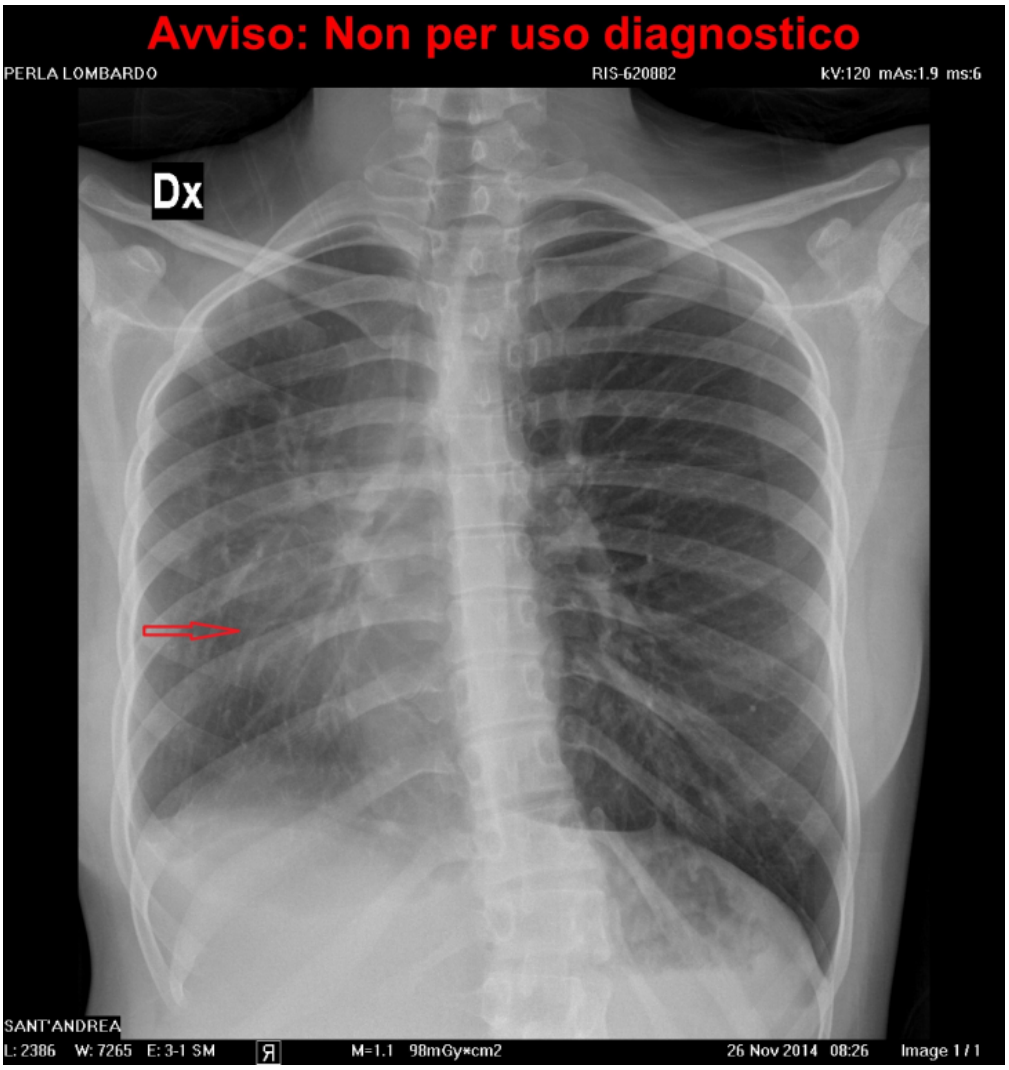
a



Description: Aplasia of the right upper lobe with consequent loss of volume of the ipsilateral lung.
Origin: Department of Radilogy, Azienda Ospedaliera Sant\'Andrea, University La Sapienza, Rome

Figure 6

a



Description: Diffuse opacity of the right lung with dextroposition of the heart. **Origin:** Department of Radiology, Azienda Ospedaliera Sant\'Andrea, University La Sapienza, Rome