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Scimitar Syndrome: About A Case Report and Review of the Literature

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Abstract **Case Report**

Scimitar syndrome or Felson's veno-lobar syndrome is a very rare condition, characterised by the association of cardiopulmonary abnormalities namely an abnormal right pulmonary venous return which is most commonly located in the inferior vena cava. We present an observation of a seven-month-old female infant who presented with acute dyspnoea. The diagnosis was suspected on chest X-ray and confirmed on CT scan which showed a single large right pulmonary vein draining into the right atrium with a scimitar pattern in sagittal section associated with dextrocardia and pulmonary sequestration. The prognosis is related to the extent of the left-right shunt and associated malformations.

Key words: Scimitar syndrome, cardiopulmonary abnormalities, thoracic and abdominal angioscan.

INTRODUCTION

Scimitar syndrome or Halasz syndrome is a rare disorder characterised by abnormal right pulmonary venous return, which may be partial or total. The right superior pulmonary vein or both right veins drain into the superior vena cava, the azygos vein, the inferior vena cava or more rarely into the right atrium.

PATIENT AND OBSERVATION

A seven-month-old female child was admitted to hospital with acute wheezing. Symptomatic treatment for bronchiolitis was initiated, but the evolution was marked by persistent respiratory distress and refusal to feed. Chest X-ray was performed and showed cardiomegaly and a right posterobasal lung opacity. Additional chest angioscan (Figure 1, Figure 2, Figure 3) showed a single large right pulmonary vein draining into the right atrium, associated with dextrocardia and cardiomegaly. There was also an associated arterial branch arising from a common trunk with the right renal artery and running to the right pulmonary base in relation to intra-lobar pulmonary sequestration (Figure 4).

Scimitar syndrome is a rare congenital anomaly characterised by partial or complete abnormal pulmonary venous drainage of the right or left lung into the inferior vena cava. It is most commonly associated with hypoplasia of the right lung, pulmonary sequestration, persistence of the left superior vena cava and cardiac dextroposition [1, 2]. Its prevalence is estimated to be between 1/100,000 and 1/33,000 live births and seems to affect mostly girls. The association between pathological pulmonary venous return and pulmonary sequestration is present in 50% of cases [3].

The search for sequestration is systematic before undertaking any surgical treatment of abnormal pulmonary venous return. Its clinical expression is highly variable, ranging from intolerance in the first days of life to incidental discovery in adults. The aberrant systemic artery most often arises from the lower part of the descending thoracic aorta or from the initial part of the abdominal aorta (as in our patient) [4].

In the majority of cases, it manifests itself in the neonatal period as congestive heart failure, usually due to pulmonary hypertension and respiratory failure [5]. Other major clinical complications are lung infections, haemoptysis and haemothorax, which are favoured by the systemic vascularity of the sequestered lung. The diagnosis is made on a frontal chest X-ray when a right basal arched opacity is seen, extending from the hilar region to the diaphragmatic dome [6].

The curative treatment of scimitar syndrome in some cases consists of major surgery involving re-implantation of the pulmonary vein into the left atrium and surgery for other possible associated cardiovascular malformations, including surgery for pulmonary sequestration and closure of an ASD [7].

CONCLUSION

Scimitar syndrome is a rare condition, still not well known, whose clinical manifestation is insidious and not very specific. Its most severe form with pulmonary sequestration rapidly leads to heart failure and recurrent respiratory infection. No treatment is necessary in asymptomatic patients; surgical treatment may be proposed in the event of complications such as a severe left-right shunt, sequestration or repeated pulmonary infections.

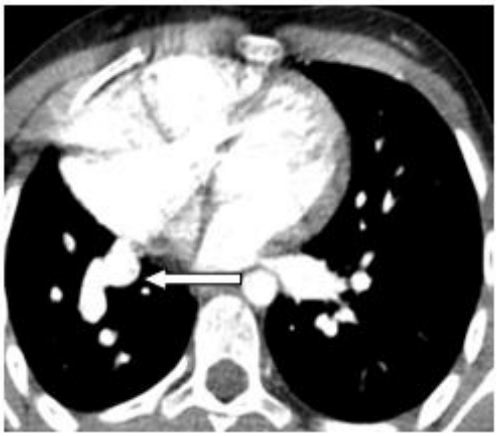

Fig-1: Axial section of thoracic angioscan: single pulmonary vein draining into the right atrium (arrow)

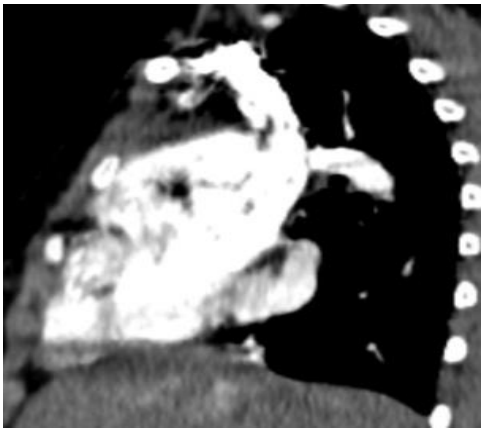

Fig-2: Sagittal section showing a single right pulmonary vein draining into the right atrium

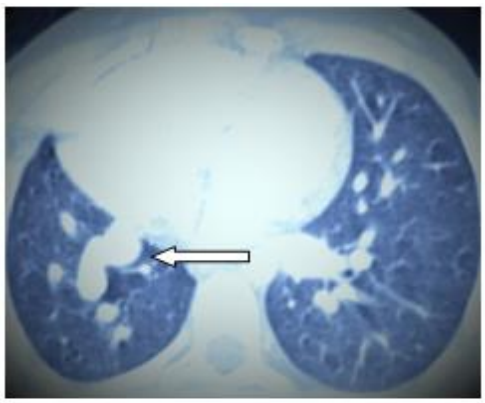

Fig-3: Axial section in parenchymal window showing right lung

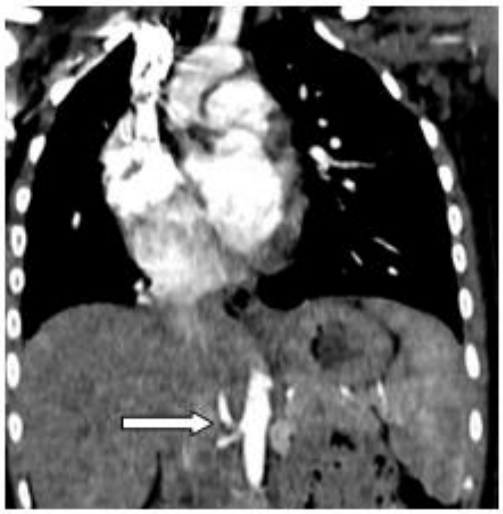

Fig-4: Feeder artery is a branch from a common trunk with the right renal artery (arrow)

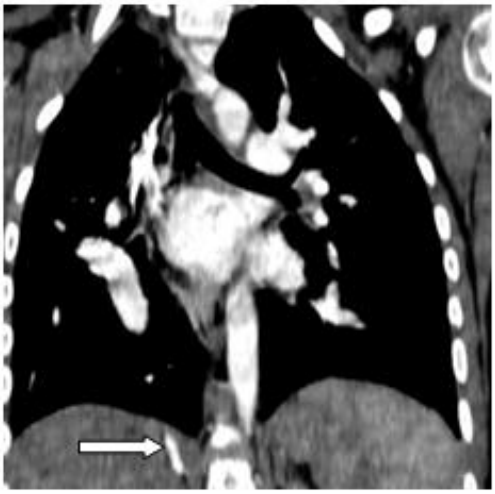

Fig-5: Coronal section of a thoracic CT scan showing the feeder artery of the right medial inferior lobar pulmonary sequestration (arrow).

RÉFÉRENCES

1. Nedelcu, C., Khalil, A., Gounant, V., Korzeck, J., Marsault, C., & Carette, M. F. (2009). Réponse du@-quid de janvier: un syndrome du cimeterre de découverte fortuite. *J Radiol*, *90*, 239-41.
2. Holt, P. D., Berdon, W. E., Marans, Z., Griffiths, S., & Hsu, D. (2004). Scimitar vein draining to the left atrium and a historical review of the scimitar syndrome. *Pediatric radiology*, *34*(5), 409-413.
3. Benjouad, I., Taam, I., Ataouna, K. E., Mahi, M., Amil, T., & Saouab, R. (2016). Scimitar syndrome: about a case and review of the literature. *The Pan African medical Journal*, *25*, 37-37.
4. Dupuis, C., Charaf, L. A., Brevière, G. M., Abou, P., Rémy-Jardin, M., & Helmius, G. (1992). The "adult" form of the scimitar syndrome. *The American journal of cardiology*, *70*(4), 502-507.
5. Carette, M. F., Frey, I., Tassart, M., Lebreton, C., & Khalil, A. (2002). Imagerie des

séquestrations. *Feuillets de radiologie*, *42*(5), 384-390.

6. Nedelcu, C., Carette, M. F., Parrot, A., Hammoudi, N., Marsault, C., & Khalil, A. (2008). Hemoptysis complicating scimitar syndrome: from diagnosis to treatment. *Cardiovascular and Interventional Radiology*, *31*(2), 96-98.
7. Kramer, U., Dörnberger, V., Fenchel, M., Stauder, N., Claussen, C. D., & Miller, S. (2003). Scimitar syndrome: morphological diagnosis and assessment of hemodynamic significance by magnetic resonance imaging. *European radiology*, *13*(6), L147-L150.