Situs Inversus With Pulmonary Atelectasis

Situs inversus asociado a atelectasia pulmonar

To the Editor,

Situs inversus totalis (SIT) is an uncommon congenital disease in which the positions of the chest and abdominal organs are reversed. Situs inversus (SI) is an autosomal recessive genetic condition that occurs in only about 0.001%–0.01% of the general population. This rare genetic anomaly is usually described or diagnosed by chance during chest or abdominal imaging procedures. We report a case of right pulmonary atelectasis in which the heart, spleen and liver were located right of midline.

A 46-year-old man, non-smoker with no particular complaints was referred to our department due to an abnormal chest X-ray showing dextrocardia and homogeneus opacity in the mid-right lung. Chest computed tomography (CT) showed segmentary atelectasis, extending from the right lower lobe to the right middle lobe (Fig. 1). Heart and spleen were also observed right of midline, while the liver was on the left. These findings were consistent with SIT. Fiberoptic bronchoscopy revealed the lack of an upper lobe in the right lung, but it is interesting to note that the structure of the upper lobe of the left lung was normal, i.e., it had 3 segments;

Fig. 1. Chest computed tomography showing soft tissue mass with attenuation coefficient for fat and irregular outline, containing a macrocalcification measuring 4 cm x 3 cm.

Conflict of Interests

We confirm that we have no conflict of interests with any constitutional government, that no pharmaceutical or medical company was involved in this report.

References

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Fig. 1. Chest computed tomography showing segmentary atelectasis extending from the right lower lobe to the right middle lobe.
apical, anterior and posterior. Bronchoalveolar lavage and cytology were normal. Bronchoalveolar lavage and aspirated fluid were negative for acid-alcohol resistant bacilli and Lowenstein culture for tuberculosis was negative. No clinical or radiological changes were observed after 6 months of follow-up.

SI is an uncommon congenital transpositional anomaly in which the organs of the abdomen develop in the wrong place. It occurs at a ratio of men to women of 3:2.1 In the normal arrangement, or situs solitus, the trilobar lung, the liver, the gall bladder and other internal organs are on the right side, but in SI they are on the left. The heart may be on the left or on the right.2

In our case, SI was defined with segmentary atelectasis of the right lung and the development of the right upper lobe in the area of the left upper lobe. To our knowledge, this combination has never been described in the worldwide literature. Sarkar et al.3 are the only authors to have reported a case of unilateral pulmonary hypoplasia associated with abdominal situs inversus in a 2-month-old infant.

In conclusion, SI is diagnosed fortuitously during a chest and abdominal imaging study. In these cases, pulmonary atelectasis in the right upper lobe of the left lung may be observed.

Conflict of Interests
The author declares no conflict of interests.

References

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Malignant Solitary Fibrous Tumour of the Pleura: An Uncommon Entity

Tumor fibroso solitario pleural maligno: una rara entidad

To the Editor,

Solitary fibrous tumor of the pleura (SFTP) is an uncommon disease, and only about 800 cases have been described in the literature. There are 2 distinct types: the local, benign form, and the more uncommon, highly aggressive diffuse form.1,2 It can appear at any age, although it is more common in patients between 60 and 70 years of age. It begins as an intrathoracic pleomorphic mass, requiring a differential diagnosis considering benign and malignant pleuropulmonary lesions. Histopathological analysis is essential for reaching the right diagnosis.

We report the case of a 55-year-old woman, non-smoker with no significant clinical history, who presented with chest pain after an injury. A chest X-ray was performed, which fortuitously revealed a right pleuropulmonary tumor. The patient had no respiratory symptoms and lung function test results were normal. A computed axial tomography (CAT) of the chest was obtained for further investigation, showing a tumor in the lower third of the right hemithorax, in close proximity to the diaphragm and cardiomedialstinal structures (Fig. 1). Fine needle biopsy revealed tissue consistent with spindle cell mesenchymal tumor. In view of these findings, the patient underwent the routine surgical procedure and the tumor was fully resected. Pathological analysis confirmed a solitary fibrous tumor with areas of spindle cells with a high mitotic index, significant pleomorphism and high cellularity (meeting the malignancy criteria of England et al.). Immunohistochemistry was positive for vimentin and CD34.3 Adjuvant treatment was ruled out due to the low chemosensitivity of this tumor strain and the lack of data in this respect. Routine monitoring was performed until 1 year later, when 2 apparently malignant nodular lesions, very close to each other (18 mm and 13 mm), were seen on a follow-up CAT. These lesions were subsequently resected. Pathological findings confirmed tumor compatible with SFTP metastasis. Six months later, multiple nodules were observed in both lungs on a repeat CAT, and bone involvement was confirmed by scintigraphy. After evaluation by the medical oncology department, c-KIT mutation determination was ordered, in view of reports of long periods of response with imatinib associated with this mutation.4 In the end, however, the tumor was c-KIT negative, so chemotherapy was started. Despite the various treatment lines administered, disease progression was rapid and the patient died shortly after the diagnosis of relapse.

This case illustrates 2 interesting aspects. Firstly, the appearance of SFTP, an uncommon tumor that presents with generalized symptoms, and must be considered as part of the differential diagnosis of pleural masses. Secondly, the disease appeared to begin as a non-aggressive type, although the histopathological finding suggested it was high risk. It subsequently gradually developed to a highly aggressive form. These findings underline that little is known about

Fig. 1. Computed axial tomography.

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