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Intrathoracic tumour

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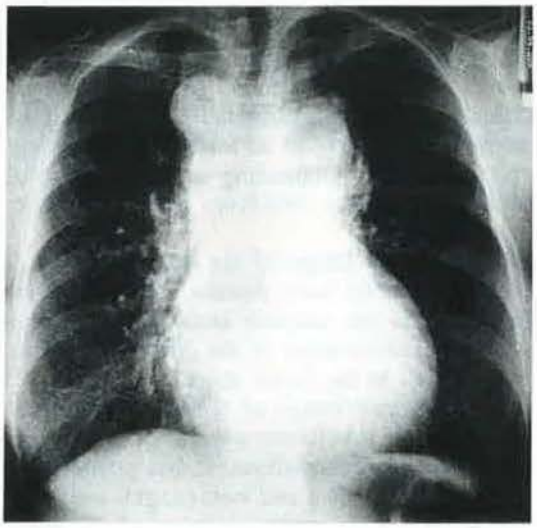
1a



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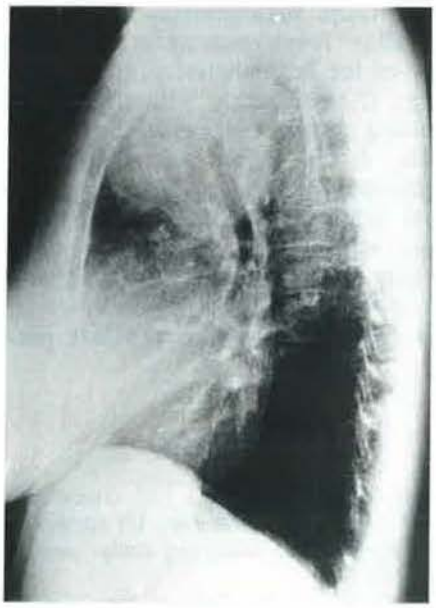

Fig. 1. – Chest roentgenograms, posteranterior (PA) and lateral views. **Interpret these before turning the page.**

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Case history

A 57 year old female was referred because of cough, hoarseness and retrosternal pain of approximately two months duration.

Indirect laryngoscopy revealed paresis of the left vocal cord. Chest roentgenograms, posteroanterior (PA) and lateral, were made (fig. 1). Bronchoscopy showed paresis of the left vocal cord, a broad main carina and irregular mucosa in the apicoposterior segmental bronchus of the left upper lobe. Histological and cytological examinations showed only inflammation without signs of malignancy.

The clinical diagnosis at this stage was a probably inoperable tumour of the left upper lobe, with an abnormal mediastinum, possibly right-sided mediastinal lymph nodes.

Magnetic resonance imaging and surgical left parasternal mediastinotomy were performed. A histological section of the tumour is shown in figure 2.

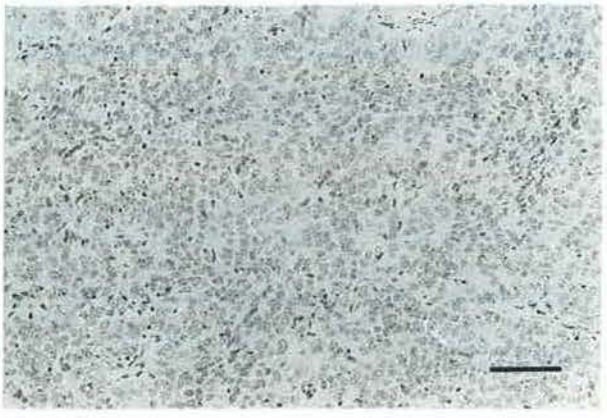

Fig. 2. – Histological section of the tumour (×140). Scale bar = 100 μm. **Suggest diagnosis before turning the page.**

Turn to next page for diagnostic tests and diagnosis

Interpretation of the chest X-ray

PA chest roentgenogram: right-sided aortic arch and tumour in the region of the left hilum.

Lateral chest roentgenogram: mass in the anterior and posterior mediastinum and hilar region.

Several weeks later an old chest and barium swallow roentgenogram became available. This confirmed the finding of a right-sided aortic arch with, on the lateral projection, a large imprint posteriorly by the left subclavian artery.

Magnetic resonance imaging (MRI)

MRI clearly showed the abnormal course of the ascending aorta and, in addition, a large cystic mass present at the left side of the superior mediastinum (fig. 3).

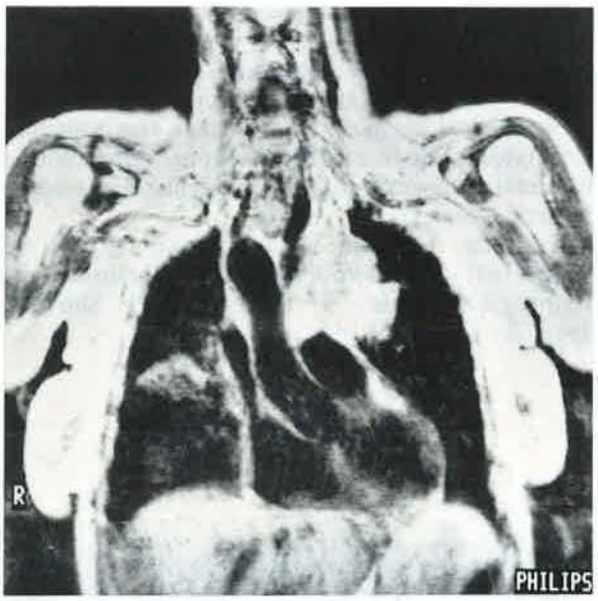

Fig. 3. – Magnetic resonance imaging (MRI) scan; right-sided aortic arch and cystic mass at the left side of the superior mediastinum.

A left parasternal mediastinotomy was performed. The histology (fig. 2) shows epithelial cells mainly located around vessels, together with reactive T-lymphocytes. The pathological diagnosis was thymoma, on clinical and histological grounds probably malignant [1, 2].

Diagnosis

Mediastinal tumour, thymoma, with a right-sided aortic arch and an aberrant left subclavian artery

Debulking was attempted by a median sternotomy. Due to the extension of the tumour and the altered anatomical situation, only 30% of the tumour was removed. Histological examination showed malignant thymoma with cortical differentiation supported by immunohistological staining for CD57 (Leu 7) and invasion of the surrounding tissue. After radiotherapy (30×2 Gy), there was some tumour regression.

Discussion

The first mention of a right-sided aortic arch came from Fioratti in 1763. In 1818, the association with the tetralogy of Fallot (Corvisart's disease) was made. Normally, in the sixth week of the foetal stage the dorsal segment of the right fourth arch obliterates. However, if the left fourth arch obliterates a right aortic arch develops. This obliteration is probably caused by haemodynamic factors as well as "controlling" tissue elements [3]. Depending on which part of the fourth arch obliterates, two types of a right aortic arch are possible [4]:

1. Type I: mirror image of the left aortic arch, associated with cyanotic heart disease. It lies anterior and to the right of the trachea and oesophagus and is caused by the obliteration of the dorsal segment of the left fourth arch in the foetal stage.
2. Type II: mirror image of the left aortic arch with an aberrant right subclavian artery, not associated with other congenital abnormalities. It lies posterior and to the right of the trachea and oesophagus and is caused by obliteration of the ventral segment of the left fourth arch.

In daily practice, it is usually sufficient to make PA and lateral chest roentgenograms together with a barium swallow roentgenogram in order to determine the origin of the left subclavian artery. Our patient had a type II aortic arch, with the left subclavian artery arising as the last vessel from the arch, an abnormality that as a rule causes no symptoms although "hesitancy in swallowing" has been reported.

The first impression of the PA chest roentgenogram was a lung tumour with an enlarged lymph node at the right side. However, this is very unlikely because a tumour in the left upper lobe usually drains through the hilar lymph nodes into the left paratracheal nodes.

References

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4. Felson B, Palayew MJ. – The two types of right aortic arch. *Radiology*, 1963; 81: 745–759.