This letter was shown to the authors who reply as follows:

Lymphomatoid granulomatosis (LYG), as described by Liebow et al.1 and subsequent reports,2,3 was defined as a distinct entity on the basis of clinicopathological features. The idea of LYG as a lymphomatous condition was suggested as early as 1977,4 and substantiated by the fact that 15% of cases were associated or evolved to lymphoma.1,2 This could not be demonstrated until late 80's by detection of clonal rearrangement of the beta T-cell receptor (b-TCR) gene.5

Our patient was diagnosed in 1985, and techniques for detection of clonal rearrangement in the b-TCR gene were not available by that time in our hospital. Therefore, the diagnosis of LYG was made on clinicopathological grounds. In our patient, only an atypical presentation delayed the diagnosis.

We chose for our patient the most suitable and efficacious treatment by the time the diagnosis was made.6 He had an acceptable quality of life up until complications developed which required surgical procedures and a long admission in an intensive care unit.

Antonio Torrelo
Manuel Martin
Antonio Rocamora
Francisco Allegue
Antonio Ledo
Department of Dermatology,
Hospital "Ramón y Cajal",
Apartado 37,
28034-Madrid,
Spain.

References


Intrapericardial phaeochromocytoma associated with two intercarotid paragangliomas: diagnostic considerations

Sir,

Intrapericardial phaeochromocytoma is rare and, as demonstrated in the following case, a challenge inasmuch as localization is concerned.

A 30 year old man with clinical and biochemical evidence of phaeochromocytoma underwent meta-iodobenzyl guanidine (MIBG) scintigraphy and a computed tomographic (CT) total body scan. These techniques were non-diagnostic and selective venous sampling was performed. This showed high plasma noradrenaline concentrations in the inferior part of the right internal jugular vein and in the right atrium. Digital subtraction angiography revealed bilateral intercarotid tumours. After surgical removal of two benign intercarotid paragangliomas, hypertension and biochemical indices of catecholamine hypersecretion were unchanged. Further MIBG scintigraphy and selective venous sampling gave similar results to those before surgery. Whereas a thoracic CT scan showed an apparently slightly enlarged left atrium and conventional two-dimensional echocardiography (2-D echo) was considered normal, a coronary angiogram revealed a mass located behind the left atrium, vascularized by the right and circumflex coronary arteries. Before operation, the lesion was differentiated from surrounding structures by magnetic resonance (MR) (performed in Grenoble) (Figure 1). At thoracotomy, an intrapericardial tumour measuring 7 × 6 × 4 cm and weighing 90 g was discovered connected to the left atrium and extending to the pulmonary vessels and the oesophagus. The resected specimen was a benign phaeochromocytoma. Now 48 months after surgery, the blood pressure and urinary levels of catecholamine and metabolites remain normal.

This case describes the difficulties encountered in the topographic diagnosis of an intrapericardial phaeochromocytoma: the tumour was associated with two non-functioning paragangliomas and was not recognized by the usual non-invasive techniques. Intrapericardial phaeochromocytoma is uncommon;1 thoracic paragangliomas account for approximately 2% of phaeochromocytoma,2 most of which are situated in the paravertebral gutter.2,3 Location in the middle mediastinum is exceptional.3

Intrapericardial paraganglioma are localized with great difficulty. Chest X-ray and conventional 2-D echo

Figure 1 Magnetic resonance imaging showing a mass (arrow) connected to the left atrium.