Retroperitoneal fibrosis of unknown origin

Sir,

Rajemiarimoelisoa et al. have recently reported in the section ‘Images in Nephrology’ an intriguing case of retroperitoneal fibrosis of unknown origin [1]. The image provided and the pathological description reminded us of similar cases with Erdheim–Chester disease, and we would like to submit this possibility to the authors. The findings of perirenal and renal infiltration, combined with major bilateral pyelocaliceal dilatation, and fibrous tissue including histiocytes suggest considering this rare condition.

Although Erdheim–Chester disease may initially present as a localized process, it is a systemic condition. Apart from retroperitoneal infiltration, other suggestive findings include symmetrical sclerosis of long bones, cutaneous xanthelasmas, central nervous system involvement mimicking intracerebral masses, and orbital, mediastinal, or pulmonary infiltration [2]. Exophthalmos and diabetes insipidus of central origin are not rare. Erdheim–Chester disease belongs to the histiocytosis spectrum. It is a non-Langerhans cell histiocytosis, characterized by xanthomatous infiltration of foamy macrophages. Here, histiocytic cells derive from the monocyte–macrophage lineage. Using the X-chromosome inactivation pattern, a recent report substantiated a clonal population in three out of five patients with Erdheim–Chester disease [3]. These data suggest that it should be regarded as a neoplastic disorder, rather than a polyclonal reactive disease.

Over the last 6 years, five patients were diagnosed with Erdheim–Chester disease and renal involvement in our department (manuscript in preparation). When present, frank dilatation of the renal tract is often impressive, as illustrated by Rajemiarimoelisoa et al. In contrast, severe renal failure is uncommon, also exemplified by their patient. Such dissociation is in sharp contrast with the degree of renal failure encountered in other varieties of retroperitoneal fibrosis with similar pyelocaliceal distension, and should make one consider the possibility of Erdheim–Chester disease. Another distinctive feature is prominent extension of the fibrotic process within the renal pelvis and throughout the renal capsule, while mid-line structures including aorta, inferior vena cava and common iliac vessels are spared. Of note, the lateral margins of the infiltration are poorly defined by CT-scan or MRI. A smoldering but unremitting course is observed in most patients. Follow-up may extend over decades. No specific treatment is yet available, and most patients are treated conservatively.

In conclusion, in the patient reported by Rajemiarimoelisoa et al. we propose to extend investigations towards (i) checking for cutaneous or eyelid deposits, (ii) performing X-ray of long bones, (iii) reviewing pathological tissue obtained at pericardotomy for xanthomatosus infiltration and (iv) performing cerebral and thoracic magnetic resonance imaging. This may allow the definitive assessment of our Erdheim–Chester hypothesis.

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