## Preserved aortic stiffness in Erdheim Chester's coated aorta: a first report

Sirs,

Erdheim-Chester disease (ECD) is a rare, non-Langerhans form of histiocytosis, in which cardiovascular involvement may be present in as much as half of the cases (1) and accounts for a significant proportion of the deaths associated with ECD (2). A "coated aorta" aspect has been well pointed out by Serratrice *et al*. (3), and has been observed in more than 20% of all ECD cases reported in the literature (2). In this study, we investigated in two ECD patients with periaortitis, carotid intima-media thickness (IMT) and aortic stiffness, two indicators of cardiovascular health frequently used as surrogate measures of sub clinic atherosclerosis (4, 5). Arterial stiffness was assessed by two independent methods, regionally via aortic pulse wave velocity (PWV) and globally via systemic arterial compliance (SAC), as previously described (6, 7).

Patient 1, a 34-year-old male, had a history of ECD for 5 years that began with a progressive paraparesis revealing a dorsal spinal cord compression by inflammatory pseudo tumour originating from a extensive periaortitis (Fig. 1) which involved the whole aorta and the left common carotid artery. A laminectomy was performed, and histology and immunohistochemistry showed characteristic signs of ECD. He had no hypertension or diabetes but had smoked tobacco for 20 years. His body mass index was 23. Patient 2, a 64-year-old female, non-smoker, with a 5-year history of hypertension, well controlled by spironolactone, had a history of ECD for 4 years, diagnosed on typical symmetric and abnormally increased labelling of the distal ends of the long bones of the lower limbs on scanning with technetium 99. Moreover, CT scan showed a periaortitis involving the abdominal aorta with an extension to the iliac arteries. Her body mass index was 26. The two patients had normal lipid and glycaemia profiles, and no carotid plaques were visualised by ultrasound scan. They were first treated with oral prednisone which was tapered and discontinued, and had taken interferon-alpha for 2 years.

Haemodynamics and large artery characteristics were within normal range; both patients presented normal brachial blood pressure, ankle-arm pressure index, IMT, SAC and aortic PWV.

This study provided the first report of mechanical properties of the aortic wall in ECD chronic periaortitis. No previous data are available in the literature for aortitis regardless to its causes. Although numerous surgery and autopsy findings pointed out a glossy aspect of the aortic wall with a periadventitial infiltration of condensed tissue, we observed similar aortic PWV and SAC than those observed in age-matched healthy non-atheromatous subjects (6, 7), indicating preserved aortic stiffness. Surgery and autopsy reports mostly concerned patients with a long time course and severe disease complicated of retroperitoneal fibrosis (3, 8) or with severe aortic atheroma (9, 10). In our patients, no atheroma of large arteries was found as suggested by the absence of aortic and carotid plaques, the normal values for carotid IMT and for ankle-arm pressure index. We hypothesised that in our two patients, aortic wall characteristics are not impaired because ECD tissue lesions likely involve an inflammation rather than a related fibrotic process, and/or atherosclerosis. The arterial wall infiltration in ECD only involves periadventitial tissue whereas atheroma is always located between the endothelium lining and the media of the arterial wall. Consequently, we could not rule out a lower impact of ECD periaortitis on arterial stiffness than the one of atherosclerosis. Several study limits should be underlined. Firstly, the study only investigated two patients, and so may not be extrapolated to other ECD patients. Secondly, PWV and SAC have only been validated to assess large artery stiffening related to the atherosclerosis process, and may not be accurate in ECD aortitis.

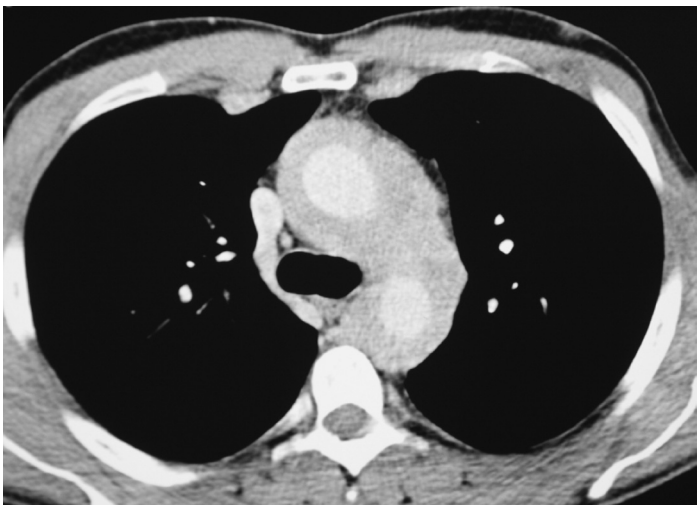

**Fig. 1.** Case 1: contrast-enhanced thoracic CT scan showing an impressive infiltration around the aortic cross.

P. ROSSI[1,3], *MD PhD*
J. SERRATRICE[2], *MD*
A. BOUSSUGES[3], *MD PhD*
Y. GOLE[3], *PhD*
A.-L. DEMOUX[1], *MD*
P.J. WEILLER[2], *MD*
B. GRANEL[1], *MD*
Y. FRANCES[1], *MD*

*[1]Department of Internal Medicine, North Hospital, Assistance Publique-Hôpitaux de Marseille (AP-HM), Marseille;*
*[2]Department of Internal Medicine, Timone Hospital, Assistance Publique-Hôpitaux de Marseille (AP-HM), Marseille;*
*[3]UMR-MD2, Physiology and Physiopathology of Extreme Oxygenation Conditions, Institut de Médecine Navale & Université Méditerranée II, Marseille, France.*

*Please address correspondence and reprint requests to: Pascal Rossi, MD PhD, Department of Internal Medicine, Assistance Publique-Hôpitaux de Marseille (AP-HM), North Hospital, Chemin des Bourrely, 13915 Marseille cedex 15, France.*
*E-mail: pascalmaurice.rossi@ap-hm.fr*

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