the findings of interstitial fibrosis, disrupted thyroid follicles, and Hürthle cells were not consistent with euthyroid Graves' disease, medically treated Graves' disease, or Hashimoto's disease, because of the lack of classic ophthalmopathy, the replacement of normal thyroid follicular epithelium by hyperplastic epithelium in less than 50% of the specimen, and the euthyroid state in this patient.⁷⁻¹⁰ Painless thyroiditis, also known as silent thyroiditis, is another possibility; this is characterised by a painless thyroid gland, raised serum concentrations of thyroid hormone, low radioactive iodine uptake, and spontaneously resolving hyperthyroidism.¹⁰ Histologically, painless thyroiditis resembles autoimmune thyroiditis, but according to Mizukami *et al*¹⁰ stromal fibrosis and Hürthle cells are rare in the former. After contacting the patient's clinician, we found that she had been receiving long term lithium treatment (over five years) for her psychiatric condition, and we were then able to arrive at the diagnosis of lithium associated autoimmune thyroiditis. However, without knowledge of the patient's clinical history, it probably would have been difficult for us to differentiate

between painless thyroiditis and lithium associated autoimmune thyroiditis. Treatment with methimazole may have partly affected the change from a hyperthyroid to a euthyroid state in this patient.

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Atypical manifestations in a patient with systemic lupus erythematosus

K Y Lam, F Cheung, L Y C Yam, C H Lee, K H Fung

Department of Pathology, University of Hong Kong, Queen Mary Hospital
K Y Lam

Department of Clinical Pathology, Pamela Youde Nethersole Eastern Hospital
F Cheung

Department of Medicine, Pamela Youde Nethersole Eastern Hospital
L Y C Yam
C H Lee

Department of Diagnostic Radiology, Pamela Youde Nethersole Eastern Hospital
K H Fung

Correspondence to:
Dr King-Yin Lam,
Department of Pathology,
Queen Mary Hospital,
Pokfulam Road, Hong Kong.

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Abstract

Systemic lupus erythematosus (SLE) is a chronic systemic inflammatory disease associated with the production of various autoantibodies and involvement of multiple organs. Necropsy findings in a 65 year old woman with SLE who had multiple aortic aneurysms and dissections, as well as other unusual manifestations, are described. The case illustrates the occurrence of and the difficulties encountered in the diagnosis of several diseases, namely aortic aneurysm, aortic dissection, acute pancreatitis, and *Penicillium marneffei* infection.

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Keywords: systemic lupus erythematosus; aneurysm; dissection.

Systemic lupus erythematosus (SLE) is a chronic systemic inflammatory disease associated with the production of various autoantibodies and involvement of multiple organs. We report the necropsy findings in a patient with

Figure 1 Axial contrast enhanced CT scan of distal thoracic aorta at level of the left ventricle. Presence of an intimal flap (arrowhead) across the aortic lumen at this section would have suggested aortic dissection. This was, however, found to be the undermined edge of a saccular aneurysm. The smaller compartment proved to be the true aortic lumen and the larger one to be the lumen of the saccular aneurysm at necropsy.

SLE who had multiple aortic aneurysms and dissections, as well as other unusual manifestations.

Case report

A 65 year old woman was admitted to hospital in June 1994 for management of longstanding hypertension, and investigation of anaemia, proteinuria, and impaired renal function. At this time, the patient's haemoglobin concentration was 66 g/l, platelet count $105 \times 10^9/l$, urea 15.5 mmol/l, and creatinine 176 $\mu\text{mol/l}$. Anti-nuclear antibody (1:2560, homogeneous pattern) and anti-double-stranded DNA were detected. A diagnosis of SLE was reached, and the patient was treated with prednisolone 30 mg daily. Azathioprine (50 mg daily initially, then 100 mg daily) was introduced after two months and prednisolone was tailed down to 20 mg daily.

Figure 2 Macroscopic appearance of the aorta at necropsy showing the presence of four saccular aneurysms in the descending aorta. A metal stent was inserted to show the dissections in the aortic wall connecting these aneurysms.

The patient presented again in April 1995 with neutropenic fever, which responded to a two week course of antibiotics. She was re-admitted to hospital in May 1995 with fever and chills. A chest x ray film revealed widened mediastinum and bilateral extensive nodular shadows. Tuberculosis was confirmed after examination of a sputum sample, and treatment was started on the second day after admission. On day 9, the patient complained of colicky abdominal pain and diarrhoea. Acute pancreatitis (amylase = 2159 IU/l) complicated by acute renal failure (urea 52.6 mmol/l and creatinine 683 $\mu\text{mol/l}$) was diagnosed on day 12. A contrast enhanced computed tomography (CT) scan was interpreted as type B aortic dissection based on the presence of intimal flap separating two contrast filled compartments (fig 1).

Despite active cardiorespiratory and renal support, the patient went into end stage, multi-organ failure. On day 17, *Penicillium marneffei* was detected on blood culture. Further active treatment was withheld and the patient died on day 18.

At necropsy, the immediate cause of death was acute pancreatitis; multiple foci of necrosis were noted in the pancreatic parenchyma and the adjacent omentum. Histologically, vasculitis was not observed in the pancreas. There was no evidence of cholelithiasis or other bile duct disorders. Patches of necrosis were seen in the lungs and hilar lymph nodes. Mycobacterial infection was confirmed in these sites by Ziehl-Neelsen stain. No signs of infection with *P. marneffei* were noted despite a specific histological search.

Four saccular aneurysms, each around 4 cm in diameter, were noted in the descending portion of the thoracic aorta (fig 2). Communications were identified between these aneurysms as the aortic wall between them was dissected. Moderate atherosclerosis was seen in the wall. There was no evidence of rupture. The features could be identified by three dimensional reconstruction of the CT scan image after correlation with the necropsy findings. Microscopic examination of the dissections and aneurysms showed cholesterol clefts, fibrosis, myxoid changes, and thrombi in the wall. Vasculitis was not identified.

Discussion

Aortic aneurysm is a rare complication in patients with SLE; only eight cases have been reported so far.¹⁻⁸ All of them were young patients (age range 26–36 years). Aortic dissections were noted in seven patients (five Daily's type A and two type B) and aortic aneurysms in two patients. One patient had a saccular thoracic aortic aneurysm and a Daily's type A dissection.² The case presented here is unusual as there were multiple aneurysms in addition to the dissections. Also, the combination of Daily's type B dissections and thoracic atherosclerotic aneurysms has never been documented in patients with SLE before.

Vasculitis was detected in the aneurysmal wall in only two patients with SLE.^{7,8} Thus, the absence of vasculitis does not preclude the

possibility that SLE was the cause of the aneurysm. Side effects of treatment with steroids should also be considered in the pathogenesis of the aneurysms. Nevertheless, the contribution of steroids to the development of the aortic lesions is difficult to assess, and aortic aneurysms in an older patient with SLE may still be incidental findings.

Aortic atherosclerotic aneurysm is often present in patients with atherosclerosis and aortic dissecting aneurysm in those with systemic hypertension. Hypertension is a fairly common finding in patients with aortic aneurysm and atherosclerosis is often present in patients with aortic dissection. However, reports describing combined aortic atherosclerotic aneurysm and dissection are rare.²⁻⁹ This may be partly related to the lack of necropsy series. Furthermore, the diagnosis of these unusual combinations is difficult, as can be seen from fig 1. The presence of four saccular aneurysms with intervening dissections is also rare and it is possible that hypertension, atherosclerosis and treatment with steroids might have contributed to their development.

Acute pancreatitis, a rare complication in patients with SLE, accounted for the abdominal pain, acute renal failure and was the cause of death in our patient. The absence of pancreatic vascular lesions in this patient suggests that factors other than vasculopathy—for example, hyperlipidaemia or autoimmunity, could be implicated in the pathogenesis.

Penicillium marneffei is a fungus which is endemic in Southeast Asia. Infection has been reported in both healthy and immunocompromised patients, and once in a patient with SLE.¹⁰ It was not surprising that *P. marneffei* was not found at necropsy in this patient because the infection may be still in the early phase and the quantity of the fungi may not be sufficient for detection in autolysed necropsy tissue.

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