with undifferentiated connective tissue disease and Sjögren’s syndrome before the development of LEMS. The diversity of autoimmune phenomena, including the recent onset of LEMS, prompted us to investigate the patient’s HLA typing. In addition to the notion that this LEMS might be non-neoplastic, the distinct combination of her HLA alleles indicated the presence of an ancestral haplotype (table 1) which has been associated with various autoimmune conditions. Although polymorphisms in HLA molecules are likely to be involved in predisposition to autoimmunity, the striking association of this haplotype might also be partly explained by linkage of disease promoting genes within the central major histocompatibility complex (MHC) region. Of note, a genetically determined high setting of TNF-α has been associated with this haplotype. Raised levels of TNF-α have also been linked to LEMS. Our patient indeed showed increased serum levels of TNF-α despite inactive disease. Thus, this central cytokine might possibly play a part in the pathogenesis of at least some of the various autoimmune phenomena seen in our patient.

In summary, this report further strengthens the link between autoimmunity to connective tissue and the nervous system, together with a common genetic susceptibility region. It also demonstrates the difficulties of differentiating muscle weakness in patients with systemic autoimmunity.

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Takayasu’s arteritis with aortic aneurysm associated with Sweet’s syndrome in childhood

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S weet’s syndrome (SS) is an acute febrile neutrophilic dermatosis, characterised by the appearance of skin lesions and fever, seldom seen in children. Three reports of SS associated with aortitis in children have been published. Takayasu’s arteritis (TA), a vasculitis affecting the aorta and its branches, is quite rare in childhood. Aortic aneurysm and congestive heart failure (CHF).

CASE REPORT
A 10 month old female patient presented with fever and crusty erythematous papules, in the trunk and limbs evolving to lax dermis (fig 1). A skin biopsy showed neutrophilic infiltrate in the dermis. Seven months later, the patient presented tachycardia and arthritis in hands, wrists, and ankles.

At admission, her general status was regular, she was pale, weighed 9.9 kg (2.5–10th centile), her length was 83 cm (25–50th centile), and she had heart rate of 160 beats/min and respiratory rate of 40/min. Blood pressure was 100/40 mm Hg in the right arm, 130/70 mm Hg in the left arm, 120/68 mm Hg in the right leg, and 98/40 mm Hg in the left leg. Peripheral pulses were wide and symmetric. Cardiac examination disclosed a diastolic murmur at the left sternal margin. Laboratory tests showed haemoglobin 91 mg/l, leucocytes 14×10³/l, erythrocyte sedimentation rate 57 mm/1st h, and a negative Mantoux test. Echocardiography showed severe aortic insufficiency (AoI) and marked dilatation of the aorta. Angioresonance showed dilatation of the ascending aorta (30 mm), aortic arch (27 mm), and descending aorta (15 mm); dilatation and stenosis in the brachiocephalic branch, common carotid, and left subclavian arteries (fig 1); and the abdominal aorta and iliac caliber were decreased, with wall irregularities.

During hospitalisation, the patient presented a decrease of left upper limb pulses and ischaemia of left hand fingers. The diagnosis of TA and SS was made. Treatment was started with intravenous gammaglobulin (2 g/kg/monthly) and intravenous pulse methylprednisolone (30 mg/kg) for 3 days, monthly, followed by oral prednisone (2 mg/kg/day), progressively decreased to 10 mg/day.

Seven months later, the patient was clinically stable, the erythrocyte sedimentation rate was 24 mm/1st h, and angioresonance showed unaltered findings in the thorax, with normalisation of abdominal aorta.

DISCUSSION
Sweet’s syndrome is an acute febrile neutrophilic dermatosis, seldom seen in infancy, characterised by fever and appearance of erythematous painful nodules, plaques, and/or
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hypertension and AoI.

Such clinical manifestations in a child. 

Autoimmune and infectious factors seem to be involved.12

rheumatic diseases, such as dermatomyositis, Sjögren's syndrome, Behçet's disease, and TA. Its cause and pathogenesis are still unknown.

This case presented SS associated with TA with severe involvement of the aorta and its branches. Three cases reports of aortitis associated with SS in children have been published."7 In all of them, skin lesions evolved to scars, characterising lax dermis. Vascular symptoms and signs may be insidious for long periods. Muster et al described the case of a 16 month old baby girl diagnosed as having SS with pulmonary involvement, who improved after receiving systemic corticotherapy. After 14 months of treatment, the patient presented ascending aorta aneurysm, AoI, and coronary stenosis, and finally died."7 The anatomopathological study showed similar histological findings in skin lesions and lesions in the aorta, leading us to question whether aortitis is part of SS. Other authors believe that vascular abnormalities found in such patients fulfil the criteria of TA.

TA is a chronic inflammatory disease, affecting the aorta and its branches. In Japan, 20% of cases were found in patients aged less than 19 and just 2% in children aged less than 10 years. The American College of Rheumatology classification criteria are applicable to the paediatric group. CHF is connected with arterial hypertension, myocarditis, pericarditis, pulmonary hypertension, and AoI. Aortic insufficiency affects 7–35% of patients with TA, being more prevalent among patients with aneurysms.4–10 The patient described presented CHF, probably secondary to arterial hypertension and AoI.

Another major problem presented by this patient was the presence of aneurysms in several aortic segments and branches. In childhood, vasculitis is an important cause of aneurysms, mainly Kawasaki disease and TA. Aneurysms have been reported in 2–33% of patients with TA, normally associated with stenosis. Aneurysms and AoI are considered to provide the worst prognosis for TA evolution.9–10

Arthralgia and/or arthritis are seen in about 33% of adult patients. Tuerlinckx et al described the case of SS in a 4 month old boy, which evolved with monarthritis of the right knee, improving after the use of systemic and intra-articular corticosteroids.9 This is the second case to present such clinical manifestations in a child.

Figure 1 (A) Lax dermis (Sweet's syndrome); (B) stenotic and aneurysmatic lesions affecting the aorta and its branches.