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The manifestation of systemic vasculitis in the central nervous system – a case report

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Abstract

We present a case of a patient with systemic vasculitis suffering – besides heart, skin and gastrointestinal lesions – from the rarely reported involvement of the central nervous system. Even though the diagnosis could not be ascertained precisely, immunosuppressive therapy led to prompt regression of symptoms including initially present neurologic manifestations.

Key words: Systemic vasculitis; Churg-Strauss syndrome; magnetic resonance; central nervous system; cyclophosphamide.

Introduction

Churg-Strauss syndrome (CSS) is a rare necrotizing small-vessel vasculitis typically characterized by asthma, eosinophilia with the presence of extravascular eosinophils, pulmonary infiltrates, paranasal sinus abnormality and neuropathy (1).

Case report

A 58-year-old woman with a history of asthma, autoimmune thyroiditis and type 2 diabetes mellitus was admitted for hospitalisation for diarrhoea, generalized weakness, disorientation and blurred vision in May 2008. Neurological examination revealed central hemiparesis, the skin displayed tiny subungual and pretibial macules. Despite the elevation of cardiac markers (troponin T 1.85 μg/l and CKMB 1.47 μkat/l) and 1 mm anterior depressions on the ECG, heart ultrasound visualized no akinetic areas and the patient negated thoracic pain. The elevation of inflammatory markers (CRP 79 mg/l, leukocyte count 14.5/nl) was associated with a massive increase in the differential count of eosinophils (69%). The examination of faeces including the tests for worms and parasites was negative. Neither brain CT scan nor cerebrospinal fluid examination did elucidate progressive hemiparesis. The suspicion of vasculitis was based upon the finding of brain MRI, which indicated the impairment of terminal and branching parts of cerebral bloodstream. The images of multiple T2 hyperintense cortical and subcortical lesions with particular affection of the occipital lobe and a small lesion in the mesencephalon and right cerebellar hemisphere are shown in Fig. 1. The suspicion of systemic vasculitis was further supported by the history of asthma, presence of eosinophilia and typical involvement of the heart, skin and gastrointestinal tract.

With a history of asthma, more than 10% of eosinophils in the differential white cell blood count and neuropathy our patient fulfilled only three out of six criteria of the American College of Rheumatology (1990) for the diagnosis of Churg-Strauss syndrome (CSS) (1). Because of severe brain affliction, the patient was treated with high-dosed IV methylprednisolone (1000 mg for 5 days, administered every other day) followed by 1mg methylprednisolone/ kg/day with gradual detraction, as recommended by EULAR for the treatment of systemic vasculitis (2). On this therapy the patient improved rapidly.

Since brain MRI, intermittent erythrocyturia and tiny subungual haematomas might also be indicative of thromboembolic aetiology of the lesions, we continued with cardiac examination. Based on the repeated lack of evidence from transesophageal echocardiography and a set of haemocultures, cardiac embolism was finally excluded as the aetiology of cerebral ischemia.

Although shank skin biopsy also revealed the signs of vasculitis, neither granulomas nor eosinophilic infiltrates (typical for CSS) could be

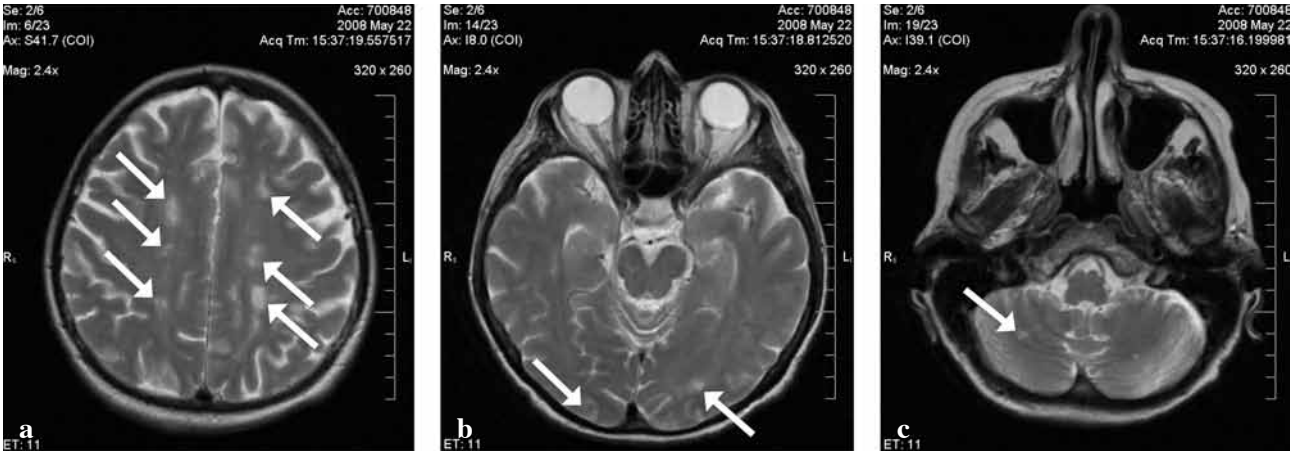

FIG. 1. — MR scan: multiple laesions in the semioval centre (a), occipital lobe and mesencephalon (b) and right cerebellar hemisphere (c).

found in the vessel wall or perivascular tissue. This led us to consider another vasculitis affecting small-sized blood vessels – a microscopic polyangiitis (MPA). Since the affection of kidneys is more frequent in MPA than in CSS (69% vs. 16%) (3), kidney biopsy was performed. However, it showed no signs of glomerulonephritis. Similarly, pANCA autoantibodies, which can be detected more often in MPA than in CSS (75% vs. 38%) (3), were absent in all blood samples. Lower respiratory tract affection was foreclosed by high resolution CT.

Owing to the high probability of generalized manifestation of systemic vasculitis the treatment with IV cyclophosphamide was commenced. This treatment is still considered fundamental for the induction of remission in generalised vasculitides (2). The patient was administered 500 mg IV cyclophosphamide monthly for 6 months. Hemiparesis, dysgraphy, impaired vision and orientation improved within 2 months. 6 months later, control MRI of the brain did not show any new lesions. Differential count and inflammatory markers were normalized.

The absence of signs of vasculitis activity enabled treatment with 2 mg azathioprine/kg/day to maintain remission (2). On this dosing regimen the patient was still in remission in August 2010.

Discussion

In the present case systemic vasculitis manifested itself in two phases typical for CSS. A prodromal phase of bronchial asthma (without detection of allergy) was succeeded by a vasculitic phase characterized by signs of multisystem disease with the affliction of the gut (diarrhoea), heart and central nervous system accompanied by eosinophilia.

Neurological manifestation of CSS is common (62-81%) (3, 4). CSS predominantly impairs the peripheral nervous system, whereas CNS involvement was documented in less than 10% of the patients (3, 4) usually in association with the affliction of other organs (5). The present patient suffered from the commonest type of CNS manifestation i.e. stroke (clinically presented as hemiparesis) and rare form i.e. confusion, vertigo, blurred vision and dysgraphy. These symptoms and signs had their correlates on MRI, e.g. blurred vision could be explained by the occipital lesion with an obvious absence of peripheral aetiology (normal eye ground, delayed visual evoked potentials).

The cardiac manifestations in CSS are highly variable. They range from (peri)myocarditis, heart failure and pericardial effusion to myocardial infarction and cardiac tamponade. Their prevalence is estimated to be 35-62% (3, 6). Even though cardiac hypokinesis could not be detected by ultrasound, cardiac involvement in our patient becomes evident as cardiospecific markers were significantly elevated and heart scan detected higher uptake of pyrophosphate.

Overall the present case illustrates that systemic vasculitis can be a risk factor for multiple cerebral infarctions necessitating prompt and adequate immunosuppressive treatment. The therapeutic efficacy does not depend on fulfilment of the full number of diagnostic criteria to ascertain the diagnosis of CNS involvement.

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