

adequately explain all the clinical features. It reflects the need for more specific criteria for diagnosing SSs.

In summary, this case highlights the importance of recognizing the very atypical presentation of sarcoidosis and its ability to mimic SSs in children, which cannot be excluded by the current classification criteria.

Rheumatology key message

- Sarcoidosis can mimic scleroderma and is not excluded by the current scleroderma classification criteria.

Disclosure statement: The authors have declared no conflicts of interest.

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Accepted 18 May 2009

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Rheumatology 2009;48:1173–1174

doi:10.1093/rheumatology/kep193

Advance Access publication 9 July 2009

Sudden visual loss in a patient with microscopic polyangiitis

SIR, A 51-year-old woman with pulmonary–renal syndrome was admitted to our department. She complained of a dry cough, which had been present for 6 months, with intermittent mild haemoptysis. One day before admission, chest CT scan suggested interstitial lung disease. At admission, blood chemistry revealed highly elevated acute phase parameters (CRP 15.6 mg/dl) and renal failure (creatinine 3.2 mg/dl). Autoantibody profiling revealed positive MPO-ANCA. Urine analysis showed granular casts, glomerular hematuria and proteinuria. Renal biopsy revealed pauci-immune crescentic glomerulonephritis. Microscopic polyangiitis (MPA) was diagnosed, and corticosteroids, cyclophosphamide and five courses of plasmapheresis commenced.

Initially, the patient responded rapidly to treatment. Inflammation markers decreased, renal function recovered and haemoptysis ceased. Despite the addition of several anti-hypertensive drugs, blood pressure, however, was rising up to 180/110 mmHg. Twenty days after immunosuppressive treatment was started, the patient suffered from severe headache and a generalized seizure. A cerebral CT scan excluded cerebral haemorrhage. Five hours later, the patient complained of complete visual loss. Cerebrospinal fluid analysis was completely unremarkable but the patient suffered from recurrent seizures necessitating transfer to an intensive care unit. MRI images showed symmetrical subcortical oedema (Fig. 1A and B). Diffusion-weighted sequences suggested cerebral ischaemia. Having excluded CNS infection, sinus vein thrombosis, subarachnoidal haemorrhage and arterial dissection, CNS vasculitis was initially suspected despite remission of pulmonary and renal disease. Methylprednisolone pulse therapy was commenced and anti-convulsant therapy started. The following day, cerebral MR angiography (MRA) showed severe bilateral narrowing of M1 and M2 cerebro-vascular segments (Fig. 1C). Over the next days, seizures subsided and the patient completely regained vision. Repeated cerebral MRA demonstrated full resolution of previously narrowed vessels and subcortical bilateral lesions (Fig. 1D–F) within 6 weeks. The patient remained in complete remission thereafter.

CNS involvement is an infrequent complication of MPA [1]. In our patient, an isolated cerebral vasculitic relapse was initially suspected after exclusion of infectious disease, sinus vein thrombosis and arterial dissection. However, the strict symmetric pattern and the rapid resolution of clinical symptoms and reversibility of arterial lesions as evidenced by MRA argue against our primary hypothesis. Also, large vessel involvement in ANCA-associated vasculitis is extremely rare. Moreover, arterial lesions of cerebral vasculitis do not usually completely resolve after healing [2]. In addition, the patient's renal and pulmonary disease responded dramatically to immunosuppressive treatment at the time of neurological deterioration. In view of these findings, we considered another diagnosis.

Recently, a group of disorders with similar clinical and angiographic findings have been described and the term reversible cerebral vasoconstriction syndrome (RCVS) has been introduced [3]. The main clinical feature of RCVS is acute onset severe headache associated with or without neurological features. The morphological basis is a prolonged but reversible multifocal cerebral vasoconstriction. Although brain MRI is frequently normal, signs of brain infarction may occur [4]. Cerebrospinal fluid analysis is normal in the majority of the cases [5]. RCVS occurs in a variety of clinical settings: drugs including cyclophosphamide, pregnancy, tumours, trauma and uncontrolled hypertension and many other factors have been reported [3]. In our patient, worsening hypertension, which occurred shortly before the clinical onset of RCVS, most likely was the final trigger, although we cannot formally exclude other causes. The optimal treatment of RCVS is uncertain but calcium-channel blockers, brief courses of corticosteroids and simple observation have been reported [6]. The previous diagnosis of systemic vasculitis in our patient complicated a correct diagnosis. To our knowledge, this is the first report of RCVS complicating ANCA-associated vasculitis.

Rheumatology key message

- Reversible cerebral vasoconstriction syndrome is an important differential diagnosis of cerebral vasculitis.

Acknowledgement

Funding: This study was supported by a research grant from the German Research Society (DFG, SFB 423, project A18).

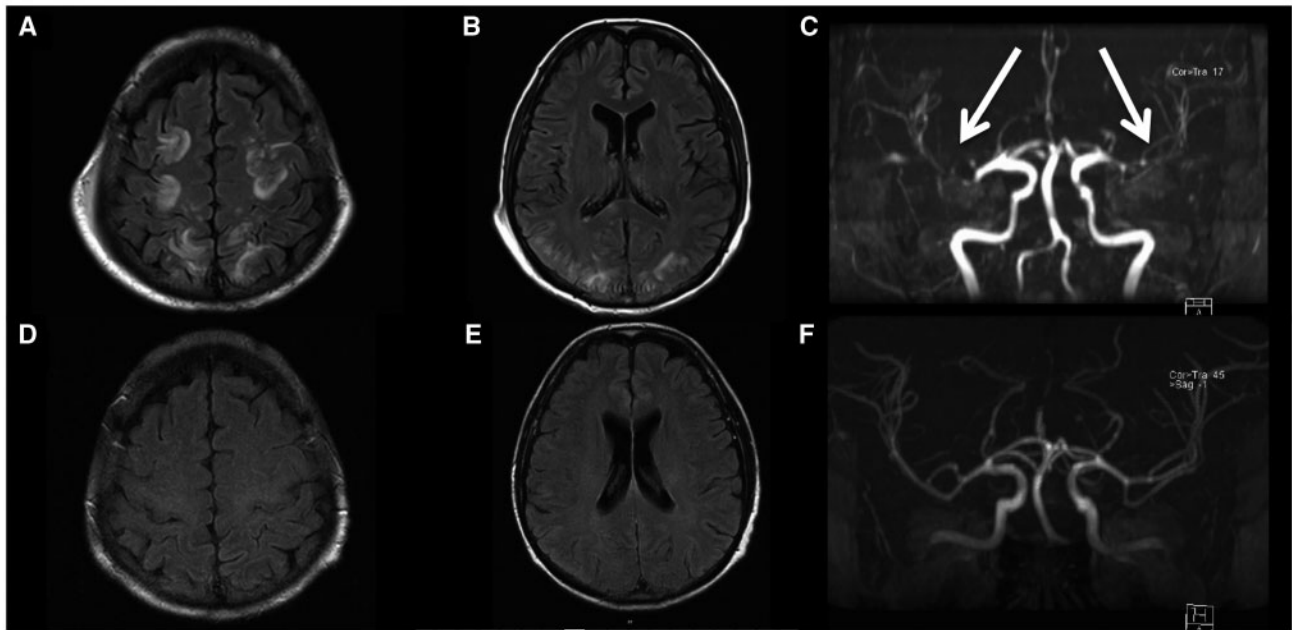


FIG. 1. Neuroimaging findings in a 51-year-old woman with MPA and reversible cerebral vasoconstriction syndrome presenting with thunderclap headache, seizures and cortical blindness. Brain MRI showed symmetrical subcortical lesions (A and B) consistent with ischaemic stroke in diffusion-weighted sequences. The following day, MRA was performed, which demonstrated severe bilateral narrowing of M1 and M2 segment arteries (C). Repeat MRA after 6 weeks revealed resolution of both subcortical lesions (D and E) and cerebral vasoconstriction (F).

Disclosure statement: The authors have declared no conflicts of interest.

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Accepted 5 June 2009

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Rheumatology 2009;48:1174–1176
doi:10.1093/rheumatology/kep210
Advance Access publication 20 July 2009

Lateral medullary syndrome with anti-neuronal antibodies (anti-Ta/Ma2) in primary Sjögren's syndrome

SIR, A 61-year-old Irish female with known primary SS, presented to the emergency department with a 3-h history of sudden-onset

vertigo, vomiting, diplopia, ataxia and hemi-facial sensory loss. She had a long-standing history of polyarthralgia and sicca symptoms with a hypergammaglobulinaemia (IgG 25g/l) and positive ANA, anti-Ro and anti-La antibodies and therefore met the diagnostic criteria for SS. Six months earlier she had an acute psychotic episode secondary to cerebral vasculitis with cerebrospinal fluid (CSF) oligoclonal bands and three small hyperintense foci in the frontal lobe on MRI of the brain. Past medical history included Grave's disease, immune thrombocytopenic purpura and coeliac disease. She admitted to poor compliance with prescribed HCQ (400 mg daily), mycophenolate mofetil (MMF) (500 mg twice daily), prednisolone (5 mg daily) and olanzapine (2.5 mg daily). On examination, she was alert and orientated. There was sensory loss to all modalities on the left side of her face and right leg, left-sided cerebellar signs (nystagmus, dysmetria, dysidiadokokinesia and truncal ataxia) and a left Horner's syndrome, consistent with a left lateral medullary syndrome.

Haematological, biochemical and inflammatory indices were within normal limits (ESR 27, CRP <5). CSF exhibited no evidence of infection. Serology demonstrated high titres of ANA, anti-Ro and anti-La antibodies. LAC, aCL and anti-dsDNA antibodies were absent. Serum western blotting for anti-neuronal antibodies was strongly positive for the anti-paraneoplastic Ma2 (PNMA2, also known as anti-Ma2/Ta) antibody only. Anti-aquaporin4 antibodies—associated with Devic's disease and recently suggested as a myelopathic association of SS—were negative [1].

MRI brain revealed two discrete foci of high signal intensity in the left medulla and left cerebellar hemisphere (Fig. 1). These were thought to be due to cerebral vasculitis rather than thromboembolism, given a normal echocardiogram and absence of aPLs. Despite the presence of anti-Ma2/Ta antibodies, a malignancy screen (CT chest, abdomen and pelvis, mammography, tumour markers, serum electrophoresis and urine Bence Jones protein) did not reveal evidence of neoplasia. There were no clinical or radiological features suggestive of lymphoma.

The patient was treated with intravenous methylprednisolone (500 mg) for 5 days and six courses of cyclophosphamide (1 g/kg