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Case Report

Churg–Strauss Syndrome Presenting as Spontaneous Subarachnoid Haemorrhage

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Abstract: Churg–Strauss syndrome (CSS) is a systemic small-vessel vasculitis characterised by the presence of asthma and eosinophilia. Central nervous system involvement (cerebral infarctions or intracerebral haemorrhage) is rare in CSS. Spontaneous subarachnoid hemorrhage (SAH) has been described in other systemic vasculitides. SAH is exceptional in CSS. We present a 47-year-old woman with CSS presenting as a spontaneous SAH with cerebral angiography findings consistent with vasculitis of the basilar artery and without aneurysms or arteriovenous malformations. She received treatment with prednisone and cyclophosphamide, and 2 months later the basilar artery was normal on magnetic resonance angiography.

Keywords: Churg–Strauss syndrome; Neurologic manifestations; Subarachnoid haemorrhage; Vasculitis

Introduction

Almost all patients with spontaneous (non-traumatic) subarachnoid haemorrhage (SAH) have ruptured saccular aneurysms or arteriovenous malformations [1]. Vasculitis is an infrequent cause of spontaneous SAH [1]. The Churg–Strauss syndrome (CSS) is a systemic small-vessel vasculitis characterised by the presence of asthma and eosinophilia [2]. The most common extrapulmonary manifestations include weight loss, fever, myalgia, arthralgia, skin signs, paranasal sinusitis, gastrointestinal tract involvement, renal disease and cardiomyopathy [2]. Neurologic involvement in CSS is also frequent, usually manifesting as peripheral neuropathy [2,3]. Central nervous system involvement (cerebral infarctions or intracerebral haemorrhage) is rare in CSS [2–4]. Spontaneous SAH has been described in other systemic vasculitides [5–7], but SAH is exceptional in CSS [8,9]. We present a 47-year-old woman with CSS presenting as a spontaneous SAH with cerebral angiography findings consisting of vasculitis of the basilar artery and without aneurysms or arteriovenous malformations.

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Case Report

A 47-year-old woman was admitted to our hospital because of severe headache. She had a long history of atopia and paranasal sinusitis. Asthma had been diagnosed 6 years earlier. There was no history of hypertension, diabetes mellitus, hyperlipidaemia, smoking or drug abuse. She had not consumed medications in the last 3 months except for inhaled fluticasone and salmeterol. Since 1 month before admission she had a purpura on the lower extremities and subsequently on the upper extremities and the trunk. She also mentioned paresthesia of the thumb, index and middle fingers of both hands and weakness of the right upper extremity. Five days before admission she developed a sudden and severe headache without nausea, vomiting or fever, which did not improve with ibuprofen.

On admission, the patient was alert and oriented. Pupils and fundi were normal. She had nuchal rigidity. Kernig's and Brudzinski's signs were negative. Pinprick sensitivity was diminished in the thumb, index and middle fingers of both hands, with weakness of wrist flexion, and there was a paralysis of the triceps and absence of the triceps reflex. Her pulse was constant at 92 beats per minute; respirations, 10 per minute; blood

pressure, 135/75 mmHg; and axillary temperature, 36.1 °C. Some wheeze was heard on lung auscultation, but heart sounds were normal with no extra sounds or murmurs. The carotid pulses were also normal without bruits. A palpable purpura was observed on the trunk and the lower and upper extremities. The remainder of the physical examination, including funduscopic examination, revealed no other abnormalities.

Remarkable blood chemical and haematologic laboratory values were: glucose 99 mg/dl, creatinine 0.7 mg/dl, total cholesterol 183 mg/dl, protein 7.2 mg/dl, fibrinogen 315 mg/dl, haemoglobin 14.4 mg/dl, platelet count 306 000/mm^3, white cell count 32 600/mm^3 (15 200 eosinophils and 12 700 neutrophils), C3 111 mg/dl (normal range 85–190 mg/dl), C4 12 mg/dl (normal range 12–36 mg/dl), albumin 43.2%, γ-globulins 22.9%, IgG 1720 mg/dl, IgM 119 mg/dl, IgA 284 mg/dl, IgE 1010 mg/dl, erythrocyte sedimentation rate 78 mm/h and C-reactive protein 13 mg/dl (normal range 0.8–5 mg/dl). Prothrombin and partial thromboplastin times were normal. Rheumatoid factor was positive. Antinuclear antibodies, antibodies to double-stranded DNA, antibodies to extractable nuclear antigen, cryoglobulins, and serology for hepatitis B and hepatitis C viruses were negative. Cytoplasmic pattern of antineutrophil cytoplasmic antibodies (ANCA) with reactivity against myeloperoxidase was detected. The urine analysis and sediment were normal. Radiographs of the chest and an electrocardiogram were normal.

On admission, a non-contrast cranial computed tomography (CT) was normal and a non-traumatic lumbar puncture yielded a cerebrospinal fluid with 160 erythrocytes/mm^3, 0 leukocytes/mm^3 and xanthocromia after centrifugation. Four-vessel cerebral angiography performed 12 h after admission revealed findings consistent with vasculitis (irregular narrowing, with typical stenoses) in the basilar artery without aneurysms, arteriovenous malformations or other abnormalities (Fig. 1). Biopsy of the palpable purpura showed necrotising vasculitis with perivascular and interstitial inflammatory infiltration (80%–90% of eosinophils) without granulomas. Electrodiagnostic examination demonstrated an axonal neuropathy of both median nerves and the right radial nerve.

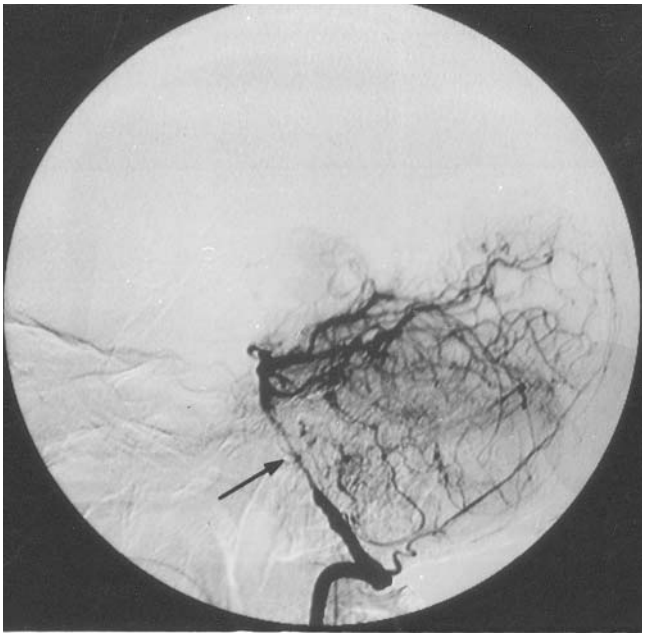

Fig. 1. Four-vessel cerebral angiography reveals findings consistent with vasculitis (irregular narrowing with typical stenoses) in the basilar artery (arrow) without aneurysms or arteriovenous malformations.

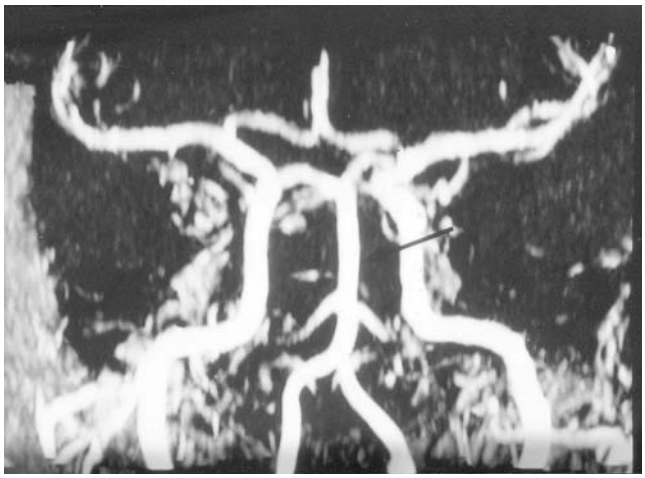

Fig. 2. Magnetic resonance angiogram of the brain shows a normal calibre of the basilar artery (arrow) after 2 months of therapy with prednisone and cyclophosphamide.

The patient received treatment with prednisone (1 mg/kg/day) and oral cyclophosphamide (2 mg/kg/day, adapted to the neutrophil count). Two months later the patient was well and a cranial magnetic resonance angiogram was normal, including the basilar artery, with normalisation of vessel calibre (Fig. 2).

Discussion

Our case had a CSS presenting as a spontaneous SAH. The American College of Rheumatology (ACR) proposed six criteria for CSS classification, with four being necessary for CSS to be diagnosed with 85% sensitivity and 99.7% specificity: asthma, eosinophilia $>10\%$, paranasal sinusitis, pulmonary infiltrate, histologic proof of vasculitis and mononeuritis multiplex [10]. Our patient presented all criteria except for the pulmonary infiltrate. The sensitivity of CT in the diagnosis of SAH decreases over time from the onset of symptoms, and a normal cranial CT is more likely in alert patients [11]. Our case presented 5 days after the onset of headache and she was alert, and cranial CT was normal. Most authorities agree that the presence of xanthocromia after centrifugation and erythrocytes in the cerebrospinal fluid, as in our case, are adequate criteria for the diagnosis of SAH in patients with normal CT [11].

Neurologic involvement in CSS is common. Peripheral neuropathy is present in 53%–78% of patients with CSS and is the second most frequent manifestation after asthma [2,3]. In contrast, central nervous system manifestations in CSS, usually consisting of cerebral infarctions, occur in only 6%–8% of cases [2,3]. Intracerebral haemorrhage has been described in CSS [4]. Other systemic vaculitides, such as Wegener's granulomatosis and isolated vasculitis of the central nervous system, have been associated with SAH [5–7]. SAH is exceptional in CSS. We have only found two such cases in a MEDLINE search for the years 1964–2000, and only one of them without aneurysms [8,9]. Angiographic demonstration of vasculitis includes irregular narrowing with typical stenoses, as in our case. Most authors accept that in the absence of tissue for biopsy the angiographic evidence of involved vessels is sufficient to make the diagnosis. Furthermore, our patient had a vasculitis confirmed by biopsy of the palpable purpura. The clinical picture, the angiographic evidence of an involved basilar artery with findings compatible with vasculitis and the demonstration of vasculitis in the skin biopsy makes it very probable that CSS was the cause of SAH in our case. We compared different techniques in the evolution, a cerebral angiography and a magnetic resonance angiogram, but the calibre of the basilar artery in the magnetic resonance angiogram after 2 months of therapy with prednisone and cyclophosphamide was completely normal.

As in our patient, asthma generally precedes the onset of the neurologic involvement [3]. The mean duration of asthma before the onset of neurologic manifestations is 6–7 years [3]. The use of coticosteroid therapy early after the onset of asthma but before that of neurologic involvement may decrease the frequency and severity of neurologic disease in CSS [3]. Central nervous system involvement may be a prognostic factor associated with a poor outcome and death in patients with CSS [2,12]. Our case highlights the need to recognise CSS as a possible but exceptional cause of SAH, especially in cases with asthma and without aneurysms or other causes of SAH.

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