headaches disappeared after 5 days. The control cerebral MRA done 40 days later demonstrated an almost complete regression of the vascular abnormalities (Fig. 1B) confirming the diagnosis of RCVS. Till today, the patient has not experienced any recurrence of ‘thunderclap headache’ after 8 months of surveillance.

True cerebral vasculitis is difficult to distinguish from RCVS on initial evaluation; in the Singhal experience [4], patients with vasculitis tend to have insidious-onset, dull headaches with stepwise clinical progression, rather than the acute, self-limited ‘thunderclap headaches’ characteristic of RCVS. Moreover, the angiographic abnormalities usually involve the distal cerebral arteries in vasculitis rather than the circle of Willis arteries or their proximal branches that are affected by vasoconstriction [4, 5]. In RCVS, spontaneous reversibility of the cerebral vasospasm within 1–3 months is the key diagnostic feature. Furthermore, cerebral vasculitis in SLE patients is rare and usually occurs in patients with active lupus [1], whereas our patient had an inactive lupus when severe headaches occurred.

Although case reports of RCVS secondary to cyclophosphamide are very rare in the literature, several authors mentioned cyclophosphamide as potential cause [3–5]. Therefore, we decided to discontinue it in our patient.

The diagnosis of RCVS in SLE patients is of practical importance, because treatment and prognosis for lupus-associated cerebral vasculitis differ strongly from those of RCVS. The clinicians must always be alerted that headaches and neurological abnormalities in patients with inactive SLE may not be CNS-SLE; awareness of the presenting clinical and neurovascular features should lead to consideration of the diagnosis of RCVS.

**Rheumatology key message**

- RCVS is an important differential diagnosis of lupus-associated cerebral vasculitis.

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**Capillaroscopic scleroderma-like pattern in patients without connective tissue disorders**

Sir, Over the past few years, there has been ever increasing interest by rheumatologists in the use of capillaroscopy, a simple and non-invasive imaging technique, due to its capability of both assessing micro-circulation abnormalities, and approaching the differential diagnosis of CTDs [1, 2]. To date, most papers indicate the scleroderma pattern as a typical marker of the scleroderma spectrum disorders (SSDs), including SSc, MCTD and dermatomyositis [1, 2].

To the best of our knowledge, this is the first report demonstrating the possibility of finding a capillaroscopic scleroderma-like pattern in conditions that are different from the so-called SSDs.

Patient 1, a 36-year-old woman, was sent to our department with suspected SSc, based on the presence of diffuse cutaneous telangiectasia. On admission, no RP or other signs or symptoms indicating a CTD were present. Physical examination revealed only numerous small telangiectasias to the face, lips, fingers and limbs. The remainder of the physical examination was remarkable. She referred to a long history of recurrent epistaxis and similar telangiectatic lesions in her sister and father. Laboratory data were normal. In particular, ANA and anti-ENA were negative. On the basis of both clinical and family history, the diagnosis of hereditary haemorrhagic telangiectasia (HHT) was made.

A nailfold videocapillaroscopy (NVC) was performed, showing clear changes in the capillary network, characterized by mega-capillaries to the third finger of the right hand, micro-haemorrhages and a diffuse enlargement of the draining limb on the remaining fingers (Fig. 1A and B).

Patient 2, a 66-year-old woman with a 15-year history of HCV infection, was admitted to our department with a long history of polyarthralgias and 4 months of lower limb paraesthesiae. She referred to episodes of purpura to the
extensor surfaces of the lower limbs, but no RP. No signs indicative of visceral involvement or other elements of SSc were found. Serology for ESR, CRP, ANA, anti-ENA, p-ANCA and c-ANCA resulted normal or negative, except for positivity for cryoglobulins. The electromyography of the lower limbs documented a mild chronic left L5/S1 radiculopathy with sensitive/motor nerve conduction velocity conserved. Thus, the diagnosis of HCV-related cryoglobulinaemia, associated with left L5/S1 radiculopathy was made. The NVC showed a scleroderma-like pattern, characterized by the presence of mega-capillaries, micro-haemorrhages, neo-angiogenesis and a diffuse enlargement of draining limbs in different fingers (Fig. 1C and D). The scleroderma pattern, which includes irregularly enlarged loops, mega-capillaries, micro-haemorrhages, neo-vascularization, loss of capillaries and architectural disorganization, has been widely documented in a variety of CTDs, particularly in SSc [1–3].

Previously, capillaroscopic features such as isolated giant capillaries have been described in patients with HHT [4, 5], but typically occurring between capillaries of normal shape and size. On the other hand, some non-specific micro-circulatory changes, including short capillaries and neo-angiogenetic phenomena have also been described in patients with mixed cryoglobulinaemia [6]. No reports describing capillaroscopic changes in patients with HCV-related cryoglobulinaemia have been published.

Our cases put in evidence an unusual presence of mega-capillaries and micro-haemorrhages associated with capillary network disarrangement, mimicking clearly a capillaroscopic scleroderma pattern.

Since the scleroderma pattern is a hallmark of SSDs, we suggest that the possibility of its presence in conditions different from the so-called SSD should be borne in mind in order to avoid misinterpretations and carry out the correct clinical approach.

**Rheumatology key message**

- Capillaroscopic scleroderma-like pattern can be found in conditions different from CTDs.

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**Fig. 1 NVC (×200 magnification).** (A) Mega-capillaries (→) and haemorrhage (arrowhead). (B) Marked enlargement of draining limbs (‘) with disordered architecture of loops. (C and D) Striking difference in loop morphology, with capillary enlargement (‘), mega-capillaries (→), haemorrhages (>) and neo-angiogenesis (○).
Unilateral glossopharyngeal and hypoglossal nerve palsies due to compression by a rheumatoid pannus

Sr., Cervical spine involvement is common in RA; however, cranial nerve palsies are comparatively rare. Here, we report a case of unilateral glossopharyngeal and hypoglossal nerve palsies related to compression by an inflammatory pannus in a patient with aggressive RA. To our knowledge, this is the first case reported where these deficits have responded to medical management.

A 46-year-old female presented with a 2-day history of sudden-onset, left-sided numbness inside her mouth and of the tongue along with difficulty in swallowing, chewing and articulating speech. There were no other sensory symptoms, weakness, headache, visual disturbances or sphincter involvement. She reported upper neck pain for several weeks, but with no recent exacerbation.

She had been diagnosed with seronegative RA 23 years previously and received a number of DMARDs including SSZ, MTX, HCO, LEF and myocrisin, in addition to corticosteroids. Grade III intraductal mammary carcinoma diagnosed in 2001 had been successfully treated surgically followed by chemotherapy and radiotherapy. Her arthritis remained poorly controlled in spite of treatment with MTX and prednisolone 10 mg daily and she underwent a number of joint replacements. In 2007, her 28-joint disease activity score (DAS-28) was 8.3 and she received two courses of rituximab, 6 months apart, with a modest initial response (DAS-28 reduction to 5.9). Rituximab was chosen in preference to anti-TNF therapy because of previous malignancy, and in keeping with the British Society for Rheumatology (BSR) guidelines [1]. However, in view of persistent active disease (DAS-28 of 8.3), a decision was made to proceed with anti-TNF therapy, in keeping with emerging clinical evidence for effectiveness and safety of this approach after rituximab [2], in accordance with the patient’s wishes, and just before her developing neurological symptoms.

At the time of presentation, examination revealed reduced sensation of the left posterior oropharynx [indicating glossopharyngeal nerve, cranial nerve (CN) IX, dysfunction] and tongue deviation to the left indicating a left hypoglossal nerve (CN XII) palsy. There was normal symmetrical movement of the uvula (i.e. normal vagus nerve, CN X, function), no dysphonia and a good cough impulse. There were no long tract signs. Active synovitis involved several joints (DAS-28 of 8.45).

Investigations showed thrombocytosis, normal biochemistry and an elevated CRP of 67 mg/l. Cervical spine X-ray revealed a marked erosion of the odontoid process and anterior subluxation of 6 mm on forward flexion. MRI showed a large inflammatory pannus measuring ~17 mm at the atlantoaxial joint, extending to the left into the region of the hypoglossal canal (Fig. 1). There was associated atlantoaxial subluxation, but no myelopathic signal change within the spinal cord. A recent mammogram and isotope bone scan had both been negative for cancer recurrence.

Treatment with i.v. methylprednisolone 1 g daily for 3 days improved her symptoms and signs. Therefore, it was felt that neurosurgical intervention was not indicated. Etanercept 50 mg s.c. once a week was commenced soon after. On clinic review after 8 weeks, the function of the glossopharyngeal nerve had returned to normal, and her swallowing and speaking difficulties had resolved. She had minimal residual tongue deviation to the left. There was a significant reduction in the DAS-28 to 4.3. This improvement was maintained at 6 months and no infections were observed. A repeat MRI scan confirmed moderate reduction in the size of pannus.

Cervical spine involvement in RA may occur in up to 80% of cases [3, 4]. Risk factors include extensive joint involvement, high seropositivity, male gender, vasculitis and prolonged use of corticosteroids [4–6]. The most common deformities affecting the cervical spine are atlantoaxial subluxation, vertical subluxation leading to superior migration of the odontoid peg (‘cranial settling’) and subaxial subluxation [3]. Compression of the spinal cord, brainstem or cranial neuropathies can result from subluxation of the spine or direct pressure by a synovial pannus. Despite the high prevalence of cervical spine involvement in RA, neurological deficits are reported in only 7–34% of patients [4, 7].

Cranial nerve palsies involving CNs IX, X and XII have been reported only rarely in patients with RA. Most of the cases have been secondary to nerve compression at the base of the skull by vertical [8, 9] or horizontal [10] subluxation at the atlantoaxial joint. Cranial neuropathy related to compression by a rheumatoid pannus has only been reported once before and involved the CN XII bilaterally [8]. Our patient had unilateral CNs IX and XII cranial neuropathy due to compression by the inflammatory pannus. To our knowledge, this is the first case reported where cranial neuropathy resolved with medical