

Localized livedo racemosa as an indicator for giant cell arteritis



Katharina Drerup, MD,^a Marcus Both, MD,^b and Thomas Schwarz, MD^a
Kiel, Germany

Key words: giant cell arteritis; immunosuppression; livedo; livedo racemosa; magnetic resonance imaging; vasculitis.

INTRODUCTION

Giant cell arteritis (GCA) represents a vasculitis of the large vessels. It can affect any organ but preferentially manifests in the temporal head area, where it is usually associated with headache, blurry vision, and necrotic ulcerations of the circulation area.¹ We here report of a patient with generalized GCA with unusual skin lesions resembling livedo racemosa.

CASE REPORT

An 85-year-old woman was admitted to a department of internal medicine because of acute abdominal pain. Acute cholecystitis was suspected due to elevated cholestasis parameters and elevated C-reactive protein, and the patient was treated with cefuroxim and metronidazole. However, the abdominal symptoms improved only moderately. The patient appeared in a weak general condition, complained about diffuse pain attacks, malaise, and fatigue. Since she developed skin lesions on the neck, she was presented to our department.

On her neck, with slight accentuation of the right side, we found bizarrely shaped, flashing figure-like, violaceous patches (Fig 1) that did not disappear on diascopy. The temples exhibited no irregular findings. The patient complained primarily about right-sided headache and scalp tenderness. In addition, she reported intermittent blurred vision in the right eye and rushing sensations in the ears. An ophthalmologic examination revealed anterior ischemic optic neuropathy. Blood tests showed leukocytosis

Abbreviations used:

GCA: giant cell arteritis
MRI: magnetic resonance imaging
SS: Sneddon syndrome



Fig 1. Bizarrely shaped, flashing figure-like, violaceous patches on the neck.

From the Department of Dermatology, University Medical Center Schleswig-Holstein, Campus Kiel^a and Department of Radiology and Neuroradiology, University Medical Center Schleswig-Holstein, Campus Kiel.^b

Funding sources: None.

IRB approval status: Not applicable.

Correspondence to: Dr Katharina Drerup, MD, Department of Dermatology, University Medical Center Schleswig-Holstein, Campus Kiel, Arnold-Heller-Straße 3, Kiel 24105, Germany. E-mail: kdrerup@dermatology.uni-kiel.de.

JAAD Case Reports 2022;24:61-3.

2352-5126

© 2022 by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jdc.2022.04.013>

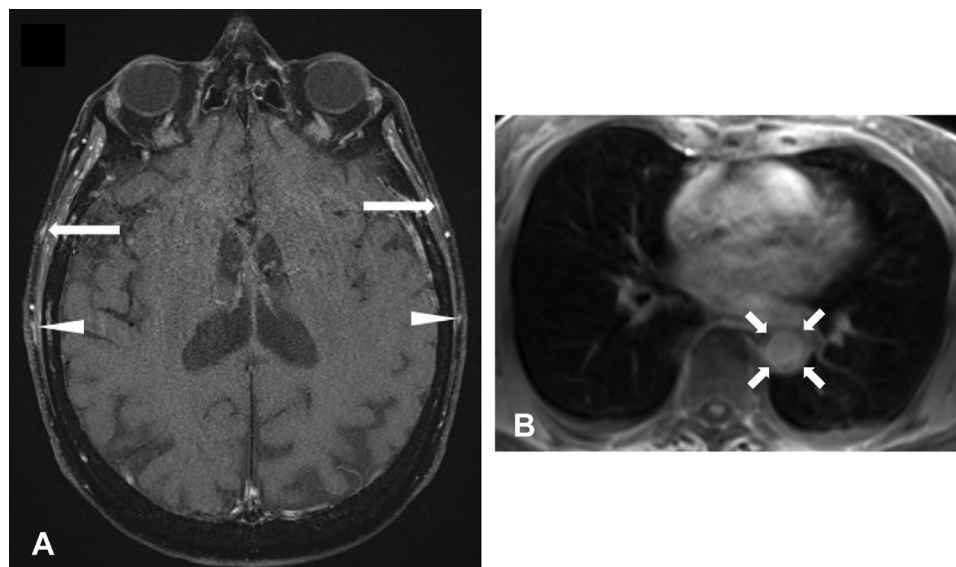


Fig 2. Cranial magnetic resonance imaging (MRI; T1-weighted, high resolution, contrast-enhanced, fat-suppressed spin-echo sequence) delineated thickening of the vessel wall and markedly increased mural and perivascular enhancement at the frontal (*arrows*) and at the parietal (*arrowheads*) branches of the superficial temporal arteries (**A**). Contrast-enhanced MRI (T1-weighted, fat-suppressed gradient-echo sequence) revealed an increased, circumferential uptake in the outer vessel wall (*arrows*) of the thoracic aorta (**B**).

($12.2 \times 10^9/L$; reference value, $3.6\text{--}10.5 \times 10^9/L$) and elevated C-reactive protein (99 mg/dL; reference value, <5 mg/dL).

Although the lesions were compatible with livedo racemosa and thus in combination with the (neurological) symptoms more indicative for Sneddon syndrome (SS), GCA was our tentative clinical diagnosis because of the half-sided headache and the episodic blurred vision. Magnetic resonance imaging (MRI) revealed a pronounced thickening of the frontal and parietal branch of the superficial temporal artery on both sides (Fig 2, A). There was also an increased accumulation of contrast media in the vessel wall of the descending aorta (Fig 2, B). Since the alterations of the temporal artery detected by MRI were highly specific for the diagnosis of active GCA, biopsy of the temporal arteries was not conducted. In turn, treatment with systemic corticosteroids (prednisolone 50 mg/day) and acetylsalicylic acid (100 mg) was initiated immediately.¹ Within 24 hours, the headache, the abdominal pain, and the general condition were significantly improved.

Upon tapering prednisolone down to 15 mg after 8 weeks, the headache and the rushing sensations in the ears reoccurred; this time accompanied by jaw claudication. Thus, the interleukin 6-receptor antagonist tocilizumab (162.5 mg/week) was administered.² Under this regimen, the patient has remained free of symptoms.

DISCUSSION

This is an unusual case for several reasons. Although the patient appeared to be severely ill, initially suffering from abdominal pain, no pathologic process could be detected. Thus, antibiotic therapy was given, as cholecystitis was suspected as the most likely cause. The skin lesions, which were reminiscent of SS, were the first indicator prompting specific investigation into neurological symptoms. SS is characterized by the appearance of livedo racemosa associated with cerebrovascular manifestations (eg, transient ischemic attacks).³ However, the neurologic examination found no evidence for a stroke attack or paresis but identified the headache as the leading symptom. Despite the lack of typical skin lesions, which usually include half-sided erosions and ulcerations on the scalp, the clinical symptoms reminded us of GCA. The cranial manifestation of GCA is typically seen in elderly patients with new-onset headache and temporal artery abnormality (tenderness to palpation or decreased pulsation) accompanied by visual impairment or loss of vision and cutaneous erosions.^{2,4,5}

In our case, the skin manifestations were not at all typical for GCA but prompted a thorough recording of the patient's history, which ultimately revealed the headache as a leading symptom. Thus, we decided for MRI of the superficial cranial arteries and of the large thoracic arteries.⁶

In the cranial MRI sequences, no cerebral infarctions or atrophy as usually present in SS could be detected. However, it turned out that the superficial temporal arteries, which are a predilection site of GCA, were affected by inflammation, along with the descending thoracic aorta as a manifestation of large-vessel vasculitis. Detailed MRI imaging of the abdominal vessels was not performed. However, since the abdominal pain rapidly disappeared upon immunosuppressive therapy, we assume that the abdominal symptoms were also caused by GCA.

Taken together, we surmise that without the appearance of the skin manifestations, the tentative diagnosis of GCA would not have been made on time; thus, the initiation of the urgently required therapy would have been delayed. It is important to mention that the skin lesions were not typical for GCA and did not yield the chance for a diagnosis at first glance; however, based on their clinical appearance and distribution, a thorough recording of the patient's history appeared relevant, which ultimately led to the correct diagnosis.⁴ Nevertheless, it remains to be mentioned that further differential diagnoses of a livedo racemosa in the facial/neck area should include

erythema *ab igne* and cholesterol emboli from the carotid arteries.⁷

Conflicts of interest

None disclosed.

REFERENCES

1. Hellmich B, Agueda A, Monti S, et al. 2018 Update of the EULAR recommendations for the management of large vessel vasculitis. *Ann Rheum Dis*. 2020;79(1):19-30. <https://doi.org/10.1136/annrheumdis-2019-215672>
2. Dechant C, Schulze-Koops H. Update Riesenzellarteriitis [Update Giant Cell Arteriitis]. *Dtsch Med Wochenschr*. 2019;144(20):1432-1436. German. <https://doi.org/10.1055/a-0722-6620>
3. Samanta D, Cobb S, Arya K. Sneddon syndrome: a comprehensive overview. *J Stroke Cerebrovasc Dis*. 2019;28(8):2098-2108. <https://doi.org/10.1016/j.jstrokecerebrovasdis.2019.05.013>
4. Lazarewicz K, Watson P. Giant cell arteritis. *BMJ*. 2019;365:l1964. <https://doi.org/10.1136/bmj.l1964>
5. McDonald L, Baker G, Kerr O. Scalp ulceration: a rare manifestation of giant cell arteritis. *BMJ Case Rep*. 2019;12(11):e230795. <https://doi.org/10.1136/bcr-2019-230795>
6. Klink T, Geiger J, Both M, et al. Giant cell arteritis: diagnostic accuracy of MR imaging of superficial cranial arteries in initial diagnosis-results from a multicenter trial. *Radiology*. 2014;273(3):844-852. <https://doi.org/10.1148/radiol.14140056>
7. Eberl M, Fink AM, Jurecka W. Facial livedo racemosa as a warning sign of cholesterol embolism from atherosclerotic plaques of the carotid artery. *J Am Acad Dermatol*. 2011;64(6):1203-1205. <https://doi.org/10.1016/j.jaad.2009.08.055>