Livedo reticularis caused by brain stem infarction: Case report and literature review

Lin Xu, Haitao Pei

ABSTRACT

Introduction: With brainstem stroke, symptoms can arise depending on the exact location of the lesion with respect to the long tracts, brainstem connection, and cranial nerve nuclei. Livedo reticularis is very characteristic for Sneddon’s syndrome. It is rarely caused by dysfunction of brain stem as described is this case. Case Report: A 50-year-old man complained of sudden onset dysarthria, dysphagia. Upon admission, neurological examination revealed unilateral weakness of the upper and lower face in the right side, Horner’s sign in the right side, severe dysphagia, and mild dysarthria, as well as insufficient temperature and pinprick sensation in left side, livedo reticularis in the extremities of the left limbs up to wrist and ankle. Brain MRI scan showed a lesion in the right medulla, which was responsible for the syndrome. Infarction in the right RVM (rostral ventral medulla) caused decreased sympathetic outflow causing livedo reticularis in the left limbs. Conclusion: In this case, Livedo reticularis in the left limbs was caused by brain stem infarction in the right, it was not Sneddon’s syndrome. This case might add discussion of other possible causes of livedo reticularis and the relevant neuroanatomy, which could explain disturbances of sympathetic nerve activity causing livedo reticularis.

Keywords: Autonomic nervous system, Brain stem stroke, Livedo reticularis, Rostral ventrolateral medulla

INTRODUCTION

Lateral medullary syndrome (Wallenberg’s syndrome) [1] is the most widely recognized brainstem syndrome. Clinical features comprise sudden-onset vertigo, vomiting, ipsilateral ataxia, ipsilateral facial numbness, ipsilateral Horner’s syndrome and contralateral loss of pain and temperature sensation in the limbs. Livedo reticularis is a dermatological disorder marked by mottled purplish discoloration of the skin due to sluggish venous blood flow [2, 3]. It is the characteristic symptom of Sneddon’s syndrome [4, 5]. It can be due to brain stem infarction rarely, as the case we will discuss here.

CASE REPORT

A 50-year-old man was admitted to our inpatient Department because of the abrupt occurrence of severe dysphagia, mild dysarthria, as well as numbness and low temperature in the left side, which meant that the sensation for temperature and pinprick was insufficient and the skin was cool to the touch. The patient had complained of mild headache on the right occipital two days before the onset. Regarding vascular risk factors, cigarette smoking was present. Upon admission,
neurological examination revealed unilateral weakness of the upper and lower face in the right side, Horner’s sign in the right side (Figure 1), severe dysphagia, and mild dysarthria, as well as insufficient temperature and pinprick sensation in left side, the skin was cool to the touch in the left side. The patient also complaints of cold sweat in the left side. Clearly we saw the livedo reticularis in the extremities of the left limbs up to wrist and ankle (Figure 2, 3). The difference of blood pressure taken in left or right was less than 15 mmHg, which meant clinically equivalent. No other neurological signs were present. He had difficulty in swallowing so we gave him nasogastric tube intubation.

A cranial CT obtained 10 hour from onset was normal. Transthoracic echocardiography was normal. Routine blood analysis and fasting lipid profile was normal, antiphospholipid antibodies, levels of antithrombin III, anti-nuclear antibodies(ANA), extractable nuclear antigen (ENA), anti-dsDNA, anti-neutrophil cytoplasmic antibodies(ANCA) were all negative or normal. The ENA included: anti-histone, anti-nRNP, anti-Sm, anti-SSA, anti- Ro-52, anti-AMA-M2, anti-SSB, anti-Rib-P, anti-Scl-70, anti-PM-Scl, anti-Jo-1, anti-centromere B, anti-PCNA, and anti-nuc as well. Brain MRI (magnetic resonance imaging, MRI) obtained one day after the onset showed a very small, diagonal lesion which has hypointensity in T1W1 and hyperintensity in DWI and T2W1, in the dorsolateral portion of the right medulla (Figures 4, 5).

Four days after onset, for better understanding of the case, we took another MRI scan. Radiographic expansion of the lesion to the lower pons in addition to the upper medulla was observed (Figures 6–8). On the fifth day after onset, the low temperature on the left arm and leg improved somewhat. And the patient had a little better speech. A thoracic spine MRI scan was taken it turned out normal (Figure 9). Computed tomography angiography (CTA) taken six days after onset revealed a severe stenosis in the right vertebral artery (Figure 10), which turned to a slight stenosis in the DSA (Digital subtraction angiography, DSA) one month later (Figure 11). We treated the patient with aspirin 300 mg per day and atorvastatin 20mg per day, as well as rehabilitation.
DISCUSSION

Livedo reticularis is a dermatological disorder marked by a mottled purplish discoloration of the skin due to sluggish venous blood flow. It is the typical symptom of Sneddon’s syndrome (SNS), but also occurs in other disorders. The diagnostic hallmarks of SNS are stroke and generalized livedo racemosa (GLR). GLR represents a...
striking violaceous, netlike patterning of the skin, similar to the familiar sign of livedo reticularis from which it differs by its shape (irregular, broken circular segments, resulting in a seemingly larger pattern), localization (both trunk and extremities), and persistence on warming. [2, 6] It generally affects young adults, with a female preponderance. Two subsets of this syndrome may be differentiated according to the absence or presence of circulating antiphospholipid (aPL) antibodies (aPL-positive SNS and aPL-negative SNS, respectively) [7]. Sneddon’s syndrome has a slow and progressive clinical course and there is no treatment that has been recognized as effective. GLR is characterized by the distribution in general in the trunk (gluteal region and inferior part of the back), from where it extends towards the extremities (along the thighs and dorsal surface of the arms). Livedo usually precedes the onset of the neurological picture, sometimes for several decades and can intensify in the acute phase of a neurological complication. The neurological picture as described by Sneddon consists of hemiplegia, aphasia, epilepsy, hemianopsia and hemianesthesia [8].

Except for SNS, there are many other causes for livedo reticularis, which could be divided into angiopathic and non-angiopathic causes. The angiopathic causes include: livedoid vasculopathy, primary antiphospholipid antibody syndrome, thromboangitis obliterans, systemic lupus erythematoses with or without antiphospholipid antibody syndrome, and rheumatic disorders; and non-angiopathic causes include: essential thrombocythaemia, polycythaemia vera, polyarthritis, pancreatitis [9]. However, the differentiation from other similar phenomenological disorders may be difficult. The evaluation for a patient with pathologic livedo reticularis is focused on determining the underlying etiology, which most commonly includes connective tissue disease,
hypercoagulable states, vasculitis, and intraluminal obstruction. Detailed differential diagnostic reflection with extensive clinical and laboratory examinations in neurological patients with livedo reticularis are therefore necessary.

As for this case, the livedo reticularis occurred simultaneously with the stroke, the distribution of Livedo reticularis was mainly in the extremities of one side, and the vessels responsible for the stroke improved dramatically in one month, so the diagnosis was not Sneddon’s syndrome, but ischemic stroke.

Considering the location of the lesion, the diagnosis could be Wallenberg syndrome[10]. Wallenberg syndrome is very common in clinic, in most cases it is due to ischemic thrombosis of posterior inferior cerebellum artery or vertebral artery. Variations of the artery make the clinical syndromes very complicated. The patient had horner’s sign on the right (Central sympathetic pathway affected), as well as hoarseness and disphagia (Nucleus ambiguous and Dorsal nucleus of the vagus nerve affected), and numbness in left limbs (Lateral spinothalamic tract affected), yet no complication of vertigo or ataxia of the contra lateral side, for neither anterior spinocerebellar tract nor inferior vestibular nucleus or inferior cerebellar peduncle was involved in the lesion. Yet the rostral ventrolateral medulla in the right was involved in this case, which would be responsible for the Livedo reticularis in the extremities of contra lateral side.

The hypothalamus is the main regulatory center for the entire peripheral autonomic system. The effenter arm of the autonomic nervous system is composed of two complementary systems, the sympathetic nervous system and the parasympathetic nervous system, whose effects are generally antagonistic to each other, yet the distinction between which is not always clear-cut. The hypothalamus exerts its regulating and controlling functions over the sympathetic and parasympathetic nervous systems by means of descending pathways which connect the hypothalamus to the descending midbrain reticular system, and in turn, carries the central impulses to the various components of the parasympathetic and sympathetic nervous systems [11]. Traditionally there is no distribution of parasympathetic nervous system in most blood vessels or sweat glands, so Livedo reticularis and sweating on the left should be due to the disinhibition of sympathetic system. Many studies demonstrated that sympathetic promoter neurons in the brainstem, at least under resting conditions, are under predominant inhibitory control from the contra lateral rostral ventrolateral medulla (RVM)[12]. Indeed, it seems the most likely conclusion in this case is that there is either 1) inhibition of sympathetic outflow to the right side of the body due to disruption of sympathetic fibers as they descend to the intermediolateral cell column in the right thoracic spinal cord [13, 14] or 2) there is decreased inhibition of sympathetic outflow to the left side of the body [15]. In the first mechanism, a global increase

in sympathetic outflow, which as noted above is well demonstrated in stroke, causes the sweating and livedo reticularis on the left side of the body. The Horner’s syndrome reported on the right face lends support to this first explanation. The two explanations are not mutually exclusive.

As indicated above, Sneddon’s syndrome is a rare clinical entity of unknown etiology characterized by the association of livedo reticularis and cerebrovascular lesions, yet there are many other diseases of potential relevance in a patient with focal cerebral ischemia may be accompanied by Livedo reticularis, due to angiopathic or non-angiopathic causes, so the differential diagnosis is very important. As for this case, Livedo reticularis was caused by disinhibition of the sympathetic system due to infarction of the contra lateral RVM.

CONCLUSION

Livedo reticularis in this patient was caused by disinhibition of the sympathetic system due to brain stem infarction. This was not a Sneddon’s syndrome.

REFERENCES


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Author Contributions
Lin Xu – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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All relevant data are within the paper and its Supporting Information files.

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