

# Lateral medullary infarction with similar features of Brown Sequard syndrome caused by vertebrobasilar dysplasia and Klippel–Feil syndrome

## A case report

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### Abstract

**Rationale:** Patients with Klippel–Feil syndrome (KFS) are always anomaly associated with vertebrobasilar dysplasia. That may present commonly as infarction of brainstem, medulla, and cerebellum. In this article, we reported a rare case of lateral medullary infarction (LMI) with similar features of Brown Sequard syndrome caused by vertebrobasilar dysplasia and KFS, and the 2 rare conditions that are causally related. The case is being reported because of its unusual and rare presentation.

**Patient concerns:** A 38-year-old female presented with acute unsteadiness, along with a tendency to lean to the left side while walking or sitting, and paresthesia in the right lower limb and trunk, at 2 days before admission. She had no history of hypertension and diabetes, but had a 20 years history of neck pain and dizziness, which was related to head movement.

**Diagnoses:** Brown Sequard syndrome and a lesion of the left thoracic spinal cord were suspected initially. KFS was confirmed by the cervical magnetic resonance imaging (MRI) and computed tomographic angiography (CTA) results. Transcranial Doppler (TCD) results confirmed that there was a causal link between LMI and KFS.

**Interventions:** The patient rejected the operation of stabilization of the cervical spine with fusion at appropriate levels.

**Outcomes:** No recurrence of stroke, but neck pain and dizziness remained after 6 months of discharge.

**Lessons:** For such patients, the conventional treatment of cerebral infarction might be ineffective, but stabilization of the cervical spine with fusion at appropriate levels can successfully prevent further episodes of syncope and stroke.

**Abbreviations:** CTA = computed tomographic angiography, DWI = diffusion-weighted imaging, EDV = end diastolic velocity, KFS = Klippel–Feil syndrome, LMI = lateral medullary infarction, MRI = magnetic resonance imaging, PSV = peak systolic velocity, TCD = transcranial Doppler.

**Keywords:** Brown Sequard syndrome, Klippel–Feil syndrome, LMI

## 1. Introduction

Klippel–Feil syndrome (KFS) was firstly described by Maurice Klippel and Andre Feil in 1912, which was a congenital spinal malformation, often accompanied by multiple fusion abnormalities

in the centrum of cervical spines, and congenital canal stenosis.<sup>[1]</sup> Spinal cord symptoms, sudden loss of consciousness, and apnea are rare neurologic complications of KFS. Cerebral infarction caused by dysplasia in vertebral artery (VA) and internal carotid artery associated with KFS had been reported.<sup>[2,3]</sup>

Lateral medullary infarction (LMI) is not uncommon in clinical practice of neurology, but in this paper, we reported a rare case of a LMI patient presented with similar features of Brown Sequard syndrome, which was caused by vertebrobasilar dysplasia and KFS.

## 2. Case report

A 38-year-old female presented with acute unsteadiness, along with a tendency to lean to the left side while walking or sitting, and paresthesia in the right lower limb and trunk, at 2 days before admission. She had no history of hypertension and diabetes, but had a 20 years history of neck pain and dizziness, which was related to head movement. And she denied these symptoms, such as double vision, slurred speech, hiccups, nausea and vomiting, dysphagia, incontinence, and facial paresthesia. Physical examination showed a short neck, low posterior hairline, and limit motion of cervical vertebra. Dysmetria was noted in left lower extremity, and there was a left side deviation when she was sitting and walking, and decreased pinprick sensation from the left

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The authors have no conflicts of interest.

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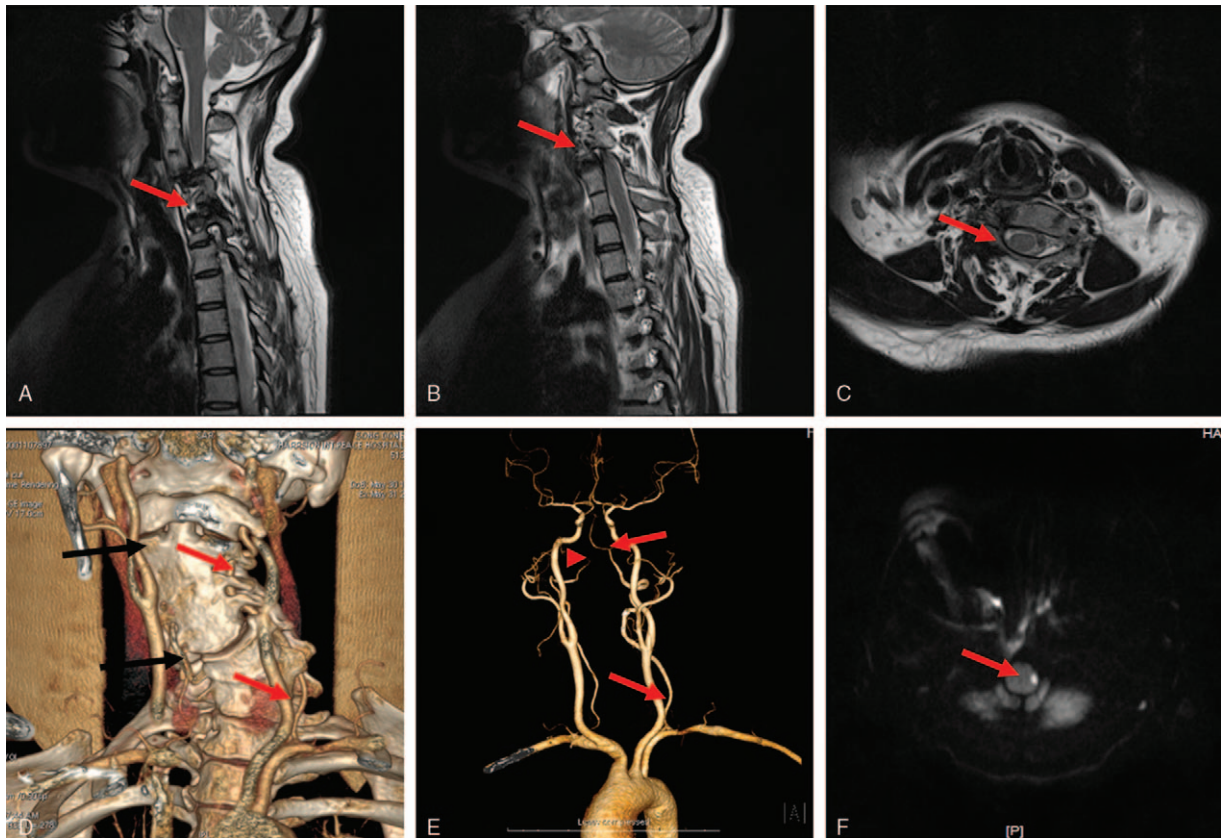
lower limb to the trunk at T4 on the right side without sacral sparing. The Romberg sign was positive but without Horner syndrome. Cranial nerves were all intact. So, Brown Sequard syndrome and a lesion of the left thoracic spinal cord were suspected initially. The thoracic magnetic resonance imaging (MRI) results were normal. On the fourth day after admission, nausea and vomiting appeared in this patient, and the physical examination showed Horner syndrome and nystagmus. Acute infarction of left lateral medulla was identified by head diffusion-weighted imaging (DWI) (Fig. 1F). Cardiac ultrasound and echocardiography were normal and no arrhythmia were confirmed. Saline foaming test was negative. The cervical MRI results revealed abnormal cervical spinal curvature and scoliosis, and partial fusion of the C2-3 and C4-5 vertebrae of cervical vertebra (Fig. 1A, B), but did not show compression of spinal cord in the C4-5 vertebrae (Fig. 1C). Computed tomographic angiography (CTA) results showed that V1 segment of right VA was slender, V2 segment was mild stenosis and did not join the basilar artery; The left VA was tortuous; bilateral fetal posterior cerebral artery; scoliosis in cervical spine and fusion malformation in vertebrae (Fig. 1D, E). Combined with the above results, KFS was confirmed. Transcranial Doppler (TCD) examination revealed peak systolic velocity (PSV) and end diastolic velocity (EDV) of the left VA, which were 45 and 18 cm/s, respectively. When her head turned left (45°), PSV and EDV of the left VA were 20 and 9 cm/s, respectively. The patient rejected the operation of stabilization of the cervical spine with fusion at appropriate

levels. Six months after discharge, there was no recurrence of stroke, but neck pain and dizziness remained.

### 3. Discussion

KFS, also known as short neck deformity, is a group of congenital malformations characterized by an obstruction in the formation and segmentation of cervical vertebrae, often associated with abnormalities of other system organs.<sup>[1]</sup> The neurological damage caused by KFS is related to compression in the spinal cord, nerve root, and blood vessels. Vascular dysplasia, such as persistence of trigeminal artery, aortic coarctation, thoracic bifurcation of the common carotid artery, and subclavian steal, is closely related to KFS.<sup>[4]</sup> At present, there are some reports of cerebral infarction caused by KFS complicated by vascular malformation and hemodynamic abnormality because of vascular compression.<sup>[2,3]</sup> In this paper, we reported a LMI patient who presented with similar features of Brown Sequard syndrome, and TCD results confirmed that there was a causal link between LMI and KFS, but both of them were relatively rare in clinic.

LMI, also known as Wallenberg syndrome, was first described in 1808 by the Genevan physician and Gaspard Vieusseux.<sup>[5]</sup> LMI is caused by obstruction of the posterior inferior cerebellar artery or VA. Its classic clinical manifestations include vertigo, nausea, vomiting, nystagmus, focal ataxia, dysphagia, dysarthria, Horner syndrome, and crossed paresthesia.<sup>[6]</sup> Our patient had no vertigo, nausea, or vomiting at the early stage, no hoarseness, dysphagia,



**Figure 1.** (A) The cervical MRI revealed partial fusion of C4-5 vertebrae of cervical vertebra (red arrow); (B) The partial fusion of C2-3 vertebrae of cervical vertebra (red arrow); (C) Cervical axial MRI (C4-5 vertebrae level) did not show compression of spinal cord (red arrow); (D, E) Computed tomographic angiography (CTA) showed cervical spine scoliosis and vertebrae fusion malformation (the part between two black arrows). The right VA did not join the basilar artery (the short arrow in E); The left VA was tortuosity in transverse foramen (Long arrow: vertebral artery); (F) The head DWI showed acute infarction of left lateral medulla (red arrow).

and no facial abnormalities and Horner sign. Paresthesia of left T4 and ataxia of left lower limb were the first manifestation. The sensory level and anatomic location of the patient should be the thoracic spinal cord. The lesion developed from the outside to inside of the spinal cord damaging thalamocortical tract, which was similar to Brown Sequard syndrome, so Brown Sequard syndrome was suspected initially. The thoracic MRI was normal, but head DWI revealed acute infarction of left lateral medulla. The patient was diagnosed with LMI, but the clinical manifestation was similar to Brown Sequard syndrome. The Brown Sequard syndrome is generally more common in spinal cord lesions; the reason why medullary infarction resembled Brown Sequard syndrome in this patient was considered to be that the arrangement of the lateral bundle from the inside to the outside of the thalamus is in the order of the neck, the thoracic, and the sacrum, and the fiber bundle in the lateral part was first involved. The reason for absence of clinical manifestations of brain stem injury at an early stage was that lesions only damaged the posterior spinocerebellar tract and the superficial thalamocortical tract, but vestibular nucleus, the descending pathway at sympathetic nervous, the spinal trigeminal tract, and the nuclei tractus spinalis nervi trigemini were relatively reserved. The patient's paresthesia was stratified at the early stage; it was related to the special hierarchical location sequence of spinothalamic tract, namely, the lumbosacral nociceptive fibers located in the lateral spinal cord, and the cervicothoracic nociceptive fibers located in the medial spinal cord. From lateral to medial spinal cord are sacral region, lower limbs, lumbar, thorax, upper limbs, and neck, respectively. Medullary blood supply was the posterior inferior cerebellar artery or VA. The pathogenesis of KFS combined with posterior circulation infarction is mostly because of the oppression of vertebral-basilar artery caused by the rotation of the head; it further damages the blood vessels, which leads to thrombosis in situ and the emboli fall off, resulting in the cerebral infarction in vertebro-basilar system.<sup>[4,7,8]</sup> Therefore, the mechanism of medullary infarction in this patient was likely to be embolic mechanism.

KFS can be associated with dysplasia of the vertebrobasilar artery,<sup>[4]</sup> and rotating the head can cause compression, damage in the blood vessels, leading to posterior circulation infarction.<sup>[9,10]</sup> And the most direct evidence is cerebral angiography or head rotation test.<sup>[9,10]</sup> Our patient was incomplete LMI and vertebrobasilar dysplasia caused by KFS; we monitored the changes of vertebrobasilar artery blood flow by TCD rotation test, and TCD revealed when she was turning head, PSV and EDV

slowed down significantly; it suggested that the pathogenesis of LMI was relative or absolute hypoperfusion of the brain stem caused by embolization of vascular injury in right VA and KFS, and this supported the causally association between LMI and KFS in this patient. In addition, the patient's history of neck pain and dizziness associated with head movement was because of the compression of the vertebral basilar artery when the head turns around; it further causes vascular injury, which leads to thrombosis in situ, and the embolus falls off and causes the vertebrobasilar artery cerebral infarction

#### 4. Conclusion

For such patients, the conventional treatment of cerebral infarction might be ineffective, but stabilization of the cervical spine with fusion at appropriate levels can successfully prevent further episodes of syncope and stroke.

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