Rubral Lateropulsion Due to Vertebral Artery Dissection in a Patient With Klippel-Feil Syndrome

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Background: Neurologic deficits in patients with Klippel-Feil syndrome usually are attributed to direct compression of neuronal structures or hypoperfusion secondary to compression of the vertebral arteries by bony abnormalities.

Objective: To describe a 38-year-old woman with known Klippel-Feil syndrome who developed lateropulsion.

Results: The results of magnetic resonance imaging were consistent with rubrothalamic stroke. The cerebral angiogram confirmed vertebral artery dissection at the level of her previously observed bony abnormality.

Conclusions: Hypermobility adjacent to fused vertebrae subjects the vertebral artery to increased shear forces. Thus, Klippel-Feil syndrome may be a predisposing factor for vertebral artery dissection. Moreover, to our knowledge, this case represents the second known case of rubral lateropulsion.

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Klippel-Feil Syndrome results from defects in segmentation predominantly affecting the posterior elements of the cervical vertebrae, leading to fusion and congenital canal stenosis.1 Neurologic complications of Klippel-Feil syndrome are infrequent, but may include spinal cord symptoms, sudden loss of consciousness, or apnea.2-6 To our knowledge, only one case report7 exists of an adult with Klippel-Feil syndrome with dissection of the vertebral artery and an associated pseudoaneurysm. Furthermore, we were able to find only one case report8 of a rubral lesion causing lateropulsion without tremor, ataxia, or weakness.

We describe a patient with 2 rare occurrences that are causally related: lateropulsion caused by rubrothalamic stroke in the context of vertebral artery dissection associated with Klippel-Feil syndrome.

REPORT OF A CASE

A 38-year-old woman was examined for bilateral hand paresthesias. She had right-sided torticollis, left-sided laterocollis, bilateral neurosensory hearing loss, short stature, and a low posterior hairline. Computed tomography of the cervical spine showed partial atlanto-occipital assimilation with fusion of the left lateral masses at C3 and C4 and left hemivertebrae at C2 and C3, confirming the diagnosis of Klippel-Feil syndrome.

One year later, she woke up with unsteadiness and a strong tendency to fall to the right. A neurologic examination revealed severe lateral sway to the right (lateropulsion), hypometric vertical saccades, gaze-evoked horizontal/torsional nystagmus accompanied by oscillopsia, and mild left-sided lower facial droop. Brain magnetic resonance imaging revealed an 8-mm acute infarct involving the left paramedian thalamus and the red nucleus (Figure 1), along with an old left cerebellar infarct. A cerebral angiogram showed left vertebral artery dissection at C2-C3 (Figure 2). The patient received aspirin, 325 mg daily. The lateropulsion and facial weakness resolved within the next week. She remained free of any new neurologic deficit over the next year.

COMMENT

Our patient presented with 2 rare conditions that are causally related: lateropulsion caused by rubrothalamic stroke, resulting from vertebral artery dissection associated with Klippel-Feil syndrome.

Neurologic complications in patients with Klippel-Feil syndrome are in-
frequent and mainly reported in children. In the older literature, deficits were attributed to mechanical compression of the brainstem without any investigation of the vertebral arteries. A vascular cause has been discussed in the literature more recently: angiographically proven compromise of vertebral arterial blood flow on extension and rotation of the head in children with Klippel-Feil syndrome has been demonstrated. In addition to compromising blood flow, hypermobility predisposes the arterial wall to shear forces during movement, causing intimal damage or dissection as a source of thromboembolism. Case reports exist of children with Klippel-Feil syndrome presenting with apnea, loss of consciousness, or posterior circulation strokes secondary to vertebral artery dissection. To our knowledge, only one case of dissection in adults has been reported. Therefore, we surmise that persisting shear forces from hypermobile vertebrae adjacent to fused segments caused the vertebral artery dissection in our patient, leading to thromboembolic stroke.

The cardinal symptom in our patient was lateropulsion, the tendency to fall laterally due to an acute uni- lateral vestibular system dysfunction in the context of normal motor function, sensation, and coordination. It is observed ipsilateral to lesions of the labyrinth, vestibular nerve, vestibular nuclei, and pontomedullary areas and contralateral to pontomesencephalic and thalamic lesions. There are case reports of ipsilateral and con-
tralateral cerebellar lesions causing lateropulsion as well. Our patient’s magnetic resonance image showed a lesion in the red nucleus and paramedian thalamus. To our knowledge, only one previous case report of rubral lateropulsion existed: the researchers postulated that the lesion in the red nucleus interrupted the ascending fibers of the crossed dentatorubrothalamic tract, causing “rubral” gait disequilibrium without tremor, ataxia, sensory loss, or weakness. Thalamic lateropulsion, also known as thalamic astasia, is usually due to a lesion in the ventrolateral thalamic area. Anatomical studies support this finding: vestibulothalamic, dentatorubrothalamic, and fastigiothalamic fibers join the thalamic fascicle adjacent to the red nucleus and end in the ventrolateral area of the thalamus (ventroposterolateral and ventrolateral). Accordingly, to our knowledge, no reports exist of paramedian thalamic lesions causing disequilibrium.

Therefore, we believe it unlikely that the paramedian thalamic lesion caused the lateropulsion in our patient. We postulate that the lesion in the red nucleus area interrupted any of the vestibulothalamic, dentatorubrothalamic, or fastigiothalamic fibers or the thalamic fascicle, providing further evidence that a lesion in the area of the red nucleus can cause lateropulsion.

Multiple researchers have already recommended flexion-extension cervical spine radiography in the examination of children presenting with vertebral artery dissection because of the many observed coexisting vertebrobasilar anomalies in such children. Because only 50% of individuals with Klippel-Feil syndrome have the classic triad of low posterior hairline, short neck, and limited cervical motion, this condition can easily escape detection.

This could theoretically lead to cases of dissection without known antecedent trauma or neck hyperextension being erroneously categorized as spontaneous. Therefore, the combination of Klippel-Feil syndrome and vertebral artery dissection may well be underreported in adults.

In conclusion, this case broadens the differential diagnosis of posterior circulation ischemic stroke, especially in young adults. We, therefore, consider it reasonable to obtain cervical spine imaging in unexplained adult cases as well. Finally, lateropulsion occurring in isolation may be secondary to a central lesion, such as in the area of the red nucleus, thus warranting neuroimaging for further examination.

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