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Gaze-Evoked and Perverted Head-Shaking Nystagmus in a Patient with Polycythemia Vera

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Polycythemia vera (PV) is well known chronic myeloproliferative neoplasm, caused by clonal expansion of an abnormal hematopoietic stem cell. Patients with PV may present diverse neurologic symptoms including headache, dizziness or vertigo, tinnitus. However, the attention has not been directed to the neuro-otological findings in patients with PV. Here, we present a 71-year-old male patient with PV suffered from vertigo and headache. He demonstrated gaze-evoked nystagmus and perverted head shaking nystagmus. Transcranial Doppler showed decrement of blood flow velocity in posterior circulation. The patient's neuro-otologic findings were normalized as polychthemia and blood flow improved with repetitive phlebotomy and medications such as hydroxyurea and aspirin. Considering the neurological and hemodynamic findings in our patient, the mechanism of vertigo in PV could be explained by central vestibulopathy because of vascular insufficiency rather than peripheral vestibulopathy because of inner ear blood hyperviscosity.

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Keywords: Nystagmus; Vertigo; Polycythemia vera; Transcranial Doppler

INTRODUCTION

Polycythemia vera (PV) is a representative chronic myelo-proliferative neoplasm caused by abnormal clonal expansion of hematopoietic stem cell [1]. Nearly 80% of patients with untreated PV have been reported neurological symptoms such as headache, dizziness, paresthesia, and chorea [2]. Among the rest, 30% of the patients with PV complain of vertigo and

dizziness [3,4]. However, the neurological findings in a dizzy patient with PV have not yet been reported. Here, we report a gaze-evoked and perverted head-shaking nystagmus with posterior hypoperfusion based on transcranial Doppler (TCD) in a patient with PV.

CASE REPORT

A 71-year-old man with PV visited our neurology department because he had suffered from episodic severe headache and vertigo for 5 days. The vertigo nature was false or distorted sensations of swaying, rocking, bobbing, or bouncing of oneself (internal non-spinning vertigo). His vertigo was accompanied by severe headache and nausea, which was aggravated by moving his head. He had no previous history

of migraine, vertigo, or episode of ataxia. He was diagnosed with PV 10 days ago by a hematology department. Vital signs were stable. On the neuro-otological examination, spontaneous nystagmus was not shown. He demonstrated bidirectional horizontal gaze-evoked and rebound nystagmus during horizontal gaze (Fig. 1). Horizontal head-shaking evoked downbeating nystagmus, i.e., perverted nystagmus (Fig. 2). Saccade was normal and smooth pursuit decreased bilaterally. Other neurological examinations were unremarkable including a cerebellar

Fig. 1. Bidirectional horizontal gaze-evoked and rebound nystagmus is shown in oculography. LH, horizontal position of the left eye; RH, horizontal position of the right eye.

Fig. 2. Perverted head shaking nystagmus is noted in oculography.

function test. A bithermal caloric test showed no caloric weakness. Pure tone audiometry showed a sensorineural hearing decrement (40 dB) in the left ear associated with a grenade injury in the Vietnam War. Brain magnetic resonance imaging and magnetic resonance angiography were unremarkable (Fig. 3). However, the TCD demonstrated the decrement of blood flow in posterior circulation (mean velocity of 13 cm/s in the basilar artery (normal range, 20–60 cm/sec), left vertebral artery (normal range, 20–50 cm/sec), 14 cm/sec in the right vertebral artery (Table 1). Serological findings were: hemoglobin of 18.8 g/100 mL, hematocrit of 62.3%, red blood cell 8,260,000/ μ L, white blood cell 16,400/ μ L, and platelets 533,000/ μ L. Biopsy samples taken from bone marrow aspiration demonstrated hypercellularity and trilineage hematopoiesis (Fig. 4). A Janus kinase 2 V617F mutation was detected. Abdominal computed tomography revealed mild

splenomegaly. The patient was treated with 1 g of hydroxyurea, 100 mg of aspirin and phlebotomy. After 3 phlebotomies and medical treatment, his symptoms and signs gradually improved with a correction of hematocrit up to 39.8% and increased blood flow on TCD (Table 1). The patient's horizontal gaze-evoked nystagmus and perverted head shaking nystagmus were disappeared simultaneously over 4 weeks.

DISCUSSION

The exact pathomechanism of vertigo in patients with PV is not well-known. Congestion of the vessels of the inner ear has been suggested as an explanation for vertigo [4]. Another possible explanation for vertigo in patients with polycythemia is intermittent insufficiency of the basilar artery [4]. We de-

Fig. 3. The patient's brain magnetic resonance imaging and magnetic resonance angiography are unremarkable.

monstrated gaze-evoked and pervverted head-shaking nystagmus in a dizzy patient with PV. Gaze-evoked nystagmus is manifest by drift of the eyes back from an eccentric position to the central position and corrective quick phases, which supports the involvement of a neural integrator that causes an

inadequately sustained eye position signal [5]. Pervverted head-shaking nystagmus is defined as nystagmus that is evoked in the plane other than that being stimulated during head-shaking (pervverted head-shaking nystagmus), i.e. downbeat nystagmus after horizontal head-shaking, which has been attributed to lesions, including in the vestibulocerebellum [6]. Therefore, the intractable vertigo in our patient supports the brainstem and cerebellar dysfunction.

Previously, reports of TCD in PV were very rare. Only a single report demonstrated the decreased blood flow, which improved after phlebotomy in PV [7]. Several previous studies observed that blood velocities of intracranial vessels were inversely proportional to hematocrit levels in healthy candidates or in patients with sickle cell anemia using TCD [8,9]. Alnaami et al. [10] previously reported the correlations of decrement of blood flow velocities and hypoperfusion of posterior circulation in patient suspected to have vertebro-basilar insufficiency. In our patient, mean flow velocities of TCD in posterior circulation were decreased at first and improved after phlebotomy and medical treatment. This blood insufficiency could give rise to a failure of neural integrator and dysfunction of vestibulocerebellum. Accordingly, considering the neurological and hemodynamic findings in our patient, the mechanism of vertigo in PV could be explained by central vestibulopathy because of vascular insufficiency rather than peripheral vestibulopathy because of inner ear blood hyperviscosity. For the first time, we documented the neuro-otological findings in a PV patient presenting with

Table 1. The blood cell profile and transcranial Doppler findings are shown before and after the phlebotomy at December 24th, 2015

Laboratory findings	2015-12-02	2015-12-09	2016-02-12
Complete blood cell counts			
Hemoglobin (g/dL)	17.4	16.9	12.9
Hematocrit (%)	58.3	56.3	38.4
Red blood cell count ($\times 10^6/\mu\text{L}$)	7.44	7.16	4.2
Mean flow velocities (cm/sec)			
Middle cerebral artery			
Left	25	24	34
Right	31	33	37
Anterior cerebral artery			
Left	23	22	49
Right	22	30	44
Posterior cerebral artery			
Left	16	18	25
Right	21	17	30
Vertebral artery			
Left	13	13	17
Right	14	14	23
Basilar artery (depth)			
7 cm	13	14	22
8 cm	15	21	34
9 cm	15	16	33
10 cm	16	20	48

Fig. 4. Trilineage proliferation is noted in peripheral blood smear (A) and bone marrow aspiration (B). H&E stain, $\times 200$.

vertigo suggesting brainstem and cerebellar dysfunction related to posterior hypoperfusion based on TCD.

중심 단어: 안진, 어지럼증, 진성적혈구증가증, 경두개초음파

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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