Original

Cerebral Hemorrhage in Turner Syndrome: A Case Report

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Abstract. We report the case of a 21-yr-old female with Turner syndrome associated with cerebral hemorrhage (CH). She was transferred to our hospital for loss of consciousness and was diagnosed with right putaminal hemorrhage. Following surgical removal of the hematoma, she regained consciousness, and her left hemiplegia gradually improved after surgery. Angiography revealed absence of vascular abnormality of the cerebral artery, aorta, and renal arteries. Hypertension was noted on arrival at the hospital and persisted after surgery. A slight hypertensive change was observed in her retinas. Plasma renin activity was elevated (20 ng/ml/h) and renovascular hypertension was suspected. In this patient, CH was suspected to have occurred due to hypertension. This case emphasizes the necessity to carefully monitor the blood pressure in Turner syndrome cases, even during childhood.

Key words: Turner syndrome, cerebral hemorrhage, hypertension

Introduction

Some cases of CH have been reported among patients with Turner syndrome (1, 2). In these cases, CH resulted from congenital vascular abnormality and hypertension. There is a relatively high prevalence of hypertension in Turner syndrome patients, compared with age-matched control groups (3–5), and most of these are renovascular hypertension cases (6, 7). Based on these findings, it is suspected that Turner syndrome patients possess some risk factors for CH. In this paper, we report a case of Turner syndrome associated with CH and discuss its causes.

Case Report

A 21-yr-old female with Turner syndrome (45,X) was transferred to our hospital due to loss of consciousness. Her consciousness level was classified as comatose (Japan Coma Scale, II-30). Left hemiplegia and hypertension (212/96 mm Hg) were observed. A computed tomography scan revealed CH in the right putaminal region (Fig. 1). Cerebral angiography revealed shifting of the right median cerebral artery due to the hematoma; however, no vascular malformation or aneurysm was found. Urinalysis, peripheral blood counts, and coagulation function tests were within the normal limits, (data not shown). Emergency surgery was performed in order to excise the hematoma, and the patient regained consciousness 12 h after the surgery. The left hemiplegia and hypertension persisted; therefore, a calcium antagonist was administered. Magnetic resonance imaging angiography revealed absence
of vascular anomaly or narrowing in the aortic arch, renal arteries, or other branches of the aorta. The optic fundi showed a slight hypertensive change, indicating that the patient was suffering from hypertension prior to this episode. Forty days after the surgery, her plasma renin activity (PRA) and plasma aldosterone concentration (PAC) were high, i.e., greater than 20.0 ng/ml/h and 331 pg/ml, respectively. Thereafter, her basal PRA persisted at a high level—from 4.0 to 10 ng/ml/h at rest in the supine position. Eight months after CH, a renin stimulation test with furosemide (20 mg i.v.) was performed with 2 h upright. Her basal renin level was 4.60 ng/ml/h, and after the stimulation, it was 34.6 ng/ml/h. Therefore, we suspected she was suffering from hyperreninemic hypertension. We administered an angiotensin II receptor blocker and a calcium antagonist, and her hypertension was well controlled. After one and a half years, her left hemiplegia was completely resolved, but the left side paresthesia persisted.

Her medical history since childhood is as follows. She was short in stature in infancy and was referred to our hospital at the age of 7 yr for evaluation of her stature (106.0 cm: –3 SD). A chromosomal study revealed the karyotype 45,X, and she was diagnosed as Turner syndrome. Recombinant human GH (rhGH) therapy was initiated from the age of 11 yr and was completed at the age of 15 yr. Following the rhGH therapy, female sex steroid replacement therapy was started. Hypertension (154/88 mm Hg) was incidentally detected at the age of 16 yr when the patient was hospitalized for a nasal conchotomy to correct a nasal obstruction. Her blood pressure, however, was not monitored thereafter. She had been living a healthy life till this cerebrovascular incident.

**Discussion**

Some cases of CH have been reported among patients with Turner syndrome (1, 2). In these cases, CH resulted from cerebral vascular abnormalities or hypertension. Turner syndrome is occasionally accompanied by aortic dissection, aortic coarctation, aneurysms, arteriovenous malformation, and telangiectasias. It is suspected that these vascular abnormalities occur due to vascular connective tissue defects (8, 9) caused by the chromosomal abnormalities in Turner syndrome. We should recognize the risk of cerebral vascular abnormality in Turner syndrome cases. The risk of CH increases if such vascular abnormalities are accompanied by hypertension.

There is a high incidence of hypertension in Turner syndrome cases in comparison with age-matched female groups (3–5). However, sex hormone replacement therapy is not recognized as elevating the blood pressure (10). With regard to hypertension, PRA is higher in Turner syndrome cases than in the control group, and it is more elevated in hypertensive Turner syndrome cases than in normotensive Turner syndrome patients (11). This high PRA may be due to the renal vascular abnormality that accompanies Turner syndrome. We should recognize the fact.
that Turner syndrome cases are susceptible to renovascular hypertension.

Our patient had both hypertension and high PRA. We could not ascertain the cause of PRA elevation, and her hypertension must have resulted from hyperreninemic hypertension. A possible mechanism of CH in our Turner syndrome patient may be the occurrence of some vascular abnormality in the renal artery and cerebral artery caused by a chromosomal abnormality. Both renovascular hypertension and cerebral vascular abnormality may play roles in the occurrence of the cerebral vascular incidents. This case emphasizes the necessity to carefully monitor the blood pressure in Turner syndrome cases, even during childhood, in order to prevent the risk of CH.

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