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Case Report

Rare Cause of Cerebellar Mutism in Childhood: Vertebral Artery Dissection

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ABSTRACT

Vertebral artery dissection (VAD), an extremely rare childhood disorder, constitutes 2%-3% of cerebrovascular diseases (CVDs). The annual incidence of CVD in childhood is estimated to be 3-8 per 100.000. Although it is generally observed after trauma, it could also be observed simultaneously with trauma. CVD is diagnosed by cranial magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA). However, the gold standard for CVD diagnosis is conventional angiography. A 12-years-old girl presenting with symptoms of headache and vomiting lasting for 5 days was hospitalized and followed up. In the neurological examination, muscle power in lower and upper extremes was 4/5 on the right and 2/5 on the left, while Babinski response was extensor on the left. Cranial MRA revealed a dissection in the right middle vertebral artery at the M1 segment, with vascular irregularity. Warfarin sodium was administered to the patient based on recommendation, and physical therapy was also started. Moderate recovery was observed in aphasia, and partial recovery was observed in left hemiplegia after 7 days of treatment. Cerebellar infarct and cerebellar mutism cases should be investigated for VAD. Early diagnosis and treatment are very pertinent for reduction of mortality and morbidity due to the disease. Angiography should be performed for diagnosis.

Keywords: Childhood, Stroke, Vertebral Artery Dissection

INTRODUCTION

The annual incidence of cerebrovascular disease (CVD) in childhood is estimated to be 3-8 per 100.000 (1). Vertebral artery dissection (VAD), an extremely rare childhood disorder, accounts for 2%-3% of CVDs (2). Since vertebral and basilar arteries are deeply located in the body, surgical intervention is often difficult, thereby posing a very high risk. Although CVD is generally observed after aneurysm, it can also occur spontaneously or due to connective tissue diseases such as Ehler-Danlos syndrome type 4 and Marfan syndrome (1, 2). VAD is generally associated with stroke due to deterioration of posterior circulation in the brain and also due to bulbar symptoms, mutism, dysarthria, vertigo, and focal findings (3). Diagnosis of VAD is determined by cranial magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA). However, the gold standard for diagnosis is conventional angiography (4). Intravascular thrombolysis, thrombectomy, and anticoagulants may be used as interventions for the treatment of VAD (5).

CASE PRESENTATION

A 12-year-old girl presenting with symptoms of headache and vomiting lasting for 5 days was hospitalized and followed-up. No anomaly was observed in the biochemical analysis and cranial computed brain tomography. After the inclusion of aphasia in the clinical symptoms, lumbar puncture revealed no cells in the cerebrospinal fluid. MRI was done in the follow-up, and the patient was directed to our clinic with suspicions of a mass in brain stem, and acute ischemia.

Cardiac apex beat was 98/min, arterial blood pressure was 104/68 mm/Hg, and fever temperature was 37^{0}C. Aphasia was observed and the patient responded appropriately to verbal signs and commands. In the neurological examination, muscle power in lower and upper extremities was 4/5 on the right and 2/5 on the left, and Babinski response was extensor on the left. The patient showed no signs of meningeal irritation while other examinations were normal. In the focal neurological examination, the patient showed a ischemic foci involving partial contrast in the cranial MRI, with bilateral diffusion limitation in thalamus and pons, which was more apparent on the right (Figure 1). Cranial MRA revealed a dissection in right middle vertebral artery at the M1 segment, with vascular irregularity (Figure 2). Enoxaparin sodium followed by warfarin sodium were administered. The patient had no history of trauma associated with the dissection. Thrombophilia panel was determined as MTHFR A1298C heterozygote and GPIIIa L33P heterozygote. Warfarin sodium was continued, while physical therapy was administered. Moderate recovery was determined in aphasia, and partial recovery was determined in left hemiplegia after 7 days of treatment. After 14 days of treatment, there was almost a total recovery in aphasia, and a moderate recovery in left hemiplegia. INR level was maintained between 2.5 and 3.5. The patient was discharged, with a

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Figure 1. Diffusion MRI images of cerebellar parenchyma, thalamus and pons where ischemic areas were observed

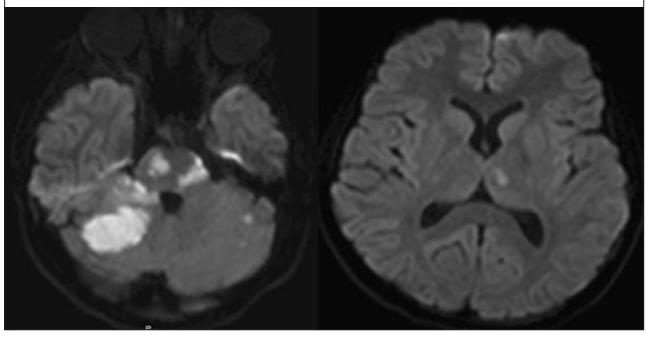
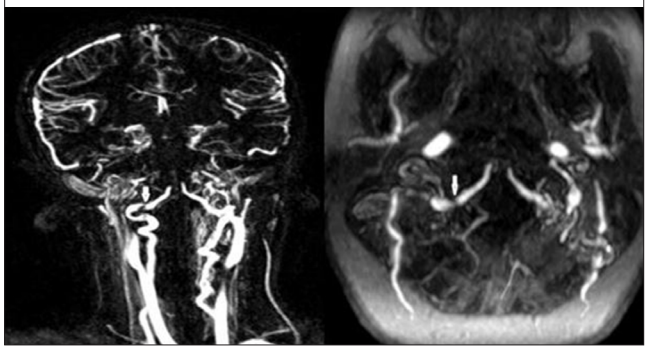


Figure 2. MR angiography, coronal and axial multiplanar reformat sequence where the vertebral artery was highly narrowed



continuation warfarin sodium treatment and regular physical therapy. After the discharge, muscle power was 5/5 on the right and 4/5 on the left after 45 days of treatment. She was walked unsupported with hemiplegic gait on the left side, with a complete recovery of her speech.

Written informed consent was obtained from the patient's family.

DISCUSSION

VAD can occur following a minor trauma or even spontaneously. It is an extremely rare disorder in childhood (1, 2). Although family history of bleeding diathesis has been detected in some cases, there is no proven pre-disposing factor for the disease (6). No trauma, syndromic findings, bleeding diathesis, and family history were observed in the present case.

Patients present different clinical symptoms based on the area affected by VAD. Patients generally present bulbar symptoms, dysarthria, vertigo, and focal symptoms (7). Although our patient had clinical symptoms of headache and vomiting, aphasia and focal symptoms were later included. Acquired neurologic childhood mutism may develop as a result of damage in different areas of the brain. Cerebellar mutism is rarely diagnosed in children. The most common reasons for the occurrence of acquired cerebellar mutism in children is the complication accompanying posterior fossa surgery (8).

Main Points:

- Cerebellar mutism is rarely diagnosed in children.
- Physical therapy is a significant method of medical treatment.
- Early diagnosis and treatment is very pertinent.

The most important imaging methods for the diagnosis of CVD are MRI and MR angiography, with conventional angiography being the gold standard method (4). Also, MRI and MRA were employed for the diagnosis of our patient since no conventional angiography was performed. There is no general consensus on the treatment of VAD at present. Current treatment interventions include administration of antithrombolitic agents and anticoagulant agents (9). We administered the anticoagulant treatment to our patient. We commenced treatment with enoxiparin sodium and warfarin sodium administrations, and later continued with warfarin sodium only. Physical therapy is an important medical treatment for morbidity in diseases caused by cerebrovascular events. We commenced physical therapy as early as possible and, as a result, obtained a positive response on the patient. Although vertebral artery dissection is a rare disorder in childhood, it poses serious threats of mortality and morbidity (6). Our patient showed better response to clinical treatment when compared to previously reported cases in literature. Complete recovery was observed in aphasia after 45 days of treatment, as well as in left hemiparesis.

CONCLUSION

Taken together, VAD is a rarely diagnosed disease in childhood. Early diagnosis and treatment is very pertinent to reduce the mortality and morbidity of the disease. VAD should be investigated in cerebellar infarct cases, and angiography should be employed for the diagnosis.

Informed Consent: Written informed consent was obtained from the patient's family.

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