Clinical Case Report

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Acute blindness as a presenting sign of left atrial myxoma in a pediatric patient

A case report and literature review

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Abstract

Rationale: Central retinal artery occlusion (CRAO) due to cardiac myxoma primarily occurs in elderly individuals. Early detection and surgical resection of myxoma are extremely important because CRAO causes complete blindness in most cases. However, due to the extremely low incidence of CRAO caused by cardiac myxoma in the pediatric age group, such condition is rarely reported.

Patient concerns: A 16-year-old female patient visited our hospital due to sudden onset of vision loss in the left eye, dysarthria, and right-sided hemiplegia.

Diagnoses: She was diagnosed with CRAO via fundoscopy. Results showed a cherry-red spot, indicating CRAO. Brain magnetic resonance imaging (MRI) revealed multifocal diffusion-restricted foci, particularly in the left frontal lobe. Echocardiography revealed a left atrial mass measuring 4.21 cm × 2.25 cm. The mass was attached to the interseptum and moved along the inflow of the mitral valve. Cardiac computed tomography (CT) revealed an enhanced mass measuring 3 cm × 2.2 cm × 3 cm and with irregular margin on the anterior wall of the left atrium and the border of the fossa ovalis.

Interventions: The patient underwent surgical excision under general anesthesia. Intraoperative finding showed a huge, jelly-like, and extremely friable mass. Pathological examination confirmed myxoma.

Outcomes: During a follow-up of 2 years after diagnosis, she did not present with other neurological deficits and no residual mass was observed on echocardiography. However, visual impairment of the left eye persisted.

Lessons: Most patients with CRAO may present with other mild symptoms that are often be neglected before CRAO development. We recommend that patients who present with frequent syncopal attack or symptoms of transient ischemic attack should undergo echocardiography.

Abbreviations: ASD = atrial septal defect, CRAO = central retinal artery occlusion, CT = computed tomography, EKG = electrocardiography, MRA = magnetic resonance angiography, MRI = magnetic resonance imaging.

Keywords: adolescent, cardiac myxoma, central retinal artery occlusion, children, pediatrics

1. Introduction

Central retinal artery occlusion (CRAO) is characterized by acute and permanent vision loss in one eye, which is a rare event, with an incidence of approximately 1 to 10 per 100,000 individuals. Such a condition is primarily observed in older adults.[1,2] In elderly individuals, CRAO is mainly caused by carotid artery atherosclerosis[3]; however, in young adults, it is typically caused by emboli arising because of cardiac disease[4–6] as well as because of cardiac myxoma. When myxoma is located in the left atrium, the risk of embolism increases because of high blood flow dynamics. In addition to embolization, individuals with myxoma in the left atrium may present with obstructive and constitutional symptoms.[7]

The embolism can occur in all body parts. However, it often causes symptoms of infarction in the middle cerebral arteries and supraclinoid internal carotid artery.[8,9] Moreover, myxoma can cause infarction in various body parts other than the brain, which include the limbs and skin, and it can also cause non–infarction-related symptoms, such as simple fatigue, syncope, and heart failure.[7,10]

Garatti et al[11] have indicated that early surgical resection of cardiac myxomas should be performed as early as possible owing to the high risk of developing systemic embolization, such as in cases of CRAO. Such a condition is associated with various

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preexisting symptoms in adults, including development of pain, occurrence of red spots on the limbs, or appearance of the transient neurologic sign.[7] However, the known characteristics of CRAO in pediatric patients are limited.

To the best of our knowledge, only 7 cases of CRAO caused by cardiac myxoma in the pediatric population have been reported thus far. Herein, we present the case of a 16-year-old female patient with CRAO caused by myxoma along with a review of literature.

2. Case presentation

A previously healthy 16-year-old girl visited the emergency department owing to sudden loss of vision in the left eye, dysarthria, numbness, and motor weakness in the right extremities. While playing on a computer, she suddenly lost her vision in the left eye and her right limbs became numb and weak. She visited the hospital the next day. She reported not paying much attention to the symptoms because she had previously experienced dysarthria and limb weakness or numbness, which had resolved quickly. However, as the symptoms persisted until the next morning, she visited the hospital.

She was diagnosed with vasovagal syncope using the tilt table test 1 year previously, and she received fludrocortisones for 4 months. At that time, the findings of brain magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), elctroencephalography, electrocardiography (EKG), and chest radiography were normal. In addition, she received dermatological care for recurrent rash on her soles. Her birth history showed no significant findings, and her growth and development was normal. There was no history of fever, headache, head injury, or seizures.

She was afebrile. No cranial or carotid bruits were noted. Cardiovascular examination revealed a regular rhythm without murmurs, gallops, or rubs. There was no hepatosplenomegaly. She was alert. However, her speech was slightly slurred. Her visual acuities in the left eye were in the category of "counting fingers." The visual field in the left eye showed a poor papillary light reflex. Neurological examination revealed right-sided motor weakness (4/5 muscle balance in the right arm and leg). Strength on the left side was normal. Muscle stretch reflexes were normal. Ophthalmic examination of the left eye revealed retinal artery attenuation and retinal whitening with a cherry-red spot in the fovea (Fig. 1). Optimal coherence tomography of the left eye revealed severe edema in the macula, and fluorescent angiography of the left eye revealed a delay in arterial filling at the early–late phase. Therefore, she was diagnosed with CRAO of the left eye. Laboratory tests revealed a white blood cell count of 10350 cells/μL, hemoglobin level of 12.5 g/dL, hematocrit level of 38.1%, and platelet count of 330,000 cells/μL. The erythrocyte sedimentation rate was 18 mm/h. Serum electrolyte, glucose, coagulation, and autoimmune test results were normal. Chest radiography revealed no cardiomegaly or pulmonary congestion. Brain diffusion-weighted MRI revealed diffusion restriction in the inferior frontal gyrus of the left frontal lobe and left thalamus, which may be consistent with acute infarction (Fig. 2). Results of brain MRA were normal. Echocardiography was performed to identify structural defects that could result in a right-to-left shunt, including the patent foramen ovale or atrial septal defect. However, echocardiography revealed a mass measuring 4.21 cm × 2.25 cm in the left atrium, which was attached to the interatrial septum. Moreover, the mass was myxomatous and movable along the inflow of the mitral valve (Fig. 3). No turbulent flow was observed. Cardiac computed tomography (CT) revealed a heterogeneously low attenuated mass in the left atrial anterior wall with an irregular margin. However, no thrombus was observed (Fig. 4). She underwent surgical excision under general anesthesia. Intraoperative finding showed a huge, jelly-like, and highly friable mass, with a pedicle on the entire septum primum. Pathological examination confirmed myxoma.

She was hospitalized for 18 days. Warfarin, enapril, and digoxin were administered for ~90, 120, and 120 days, respectively. Thereafter, treatment with aspirin was initiated.

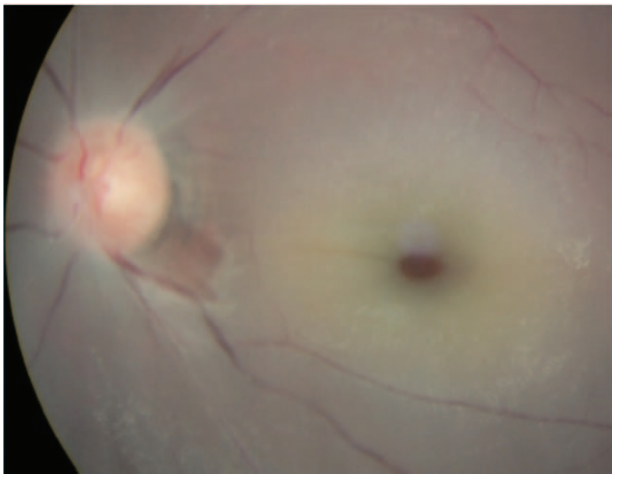

Figure 1. Fundoscopy of the left eye revealed a cherry-red spot, indicating central retinal artery occlusion.

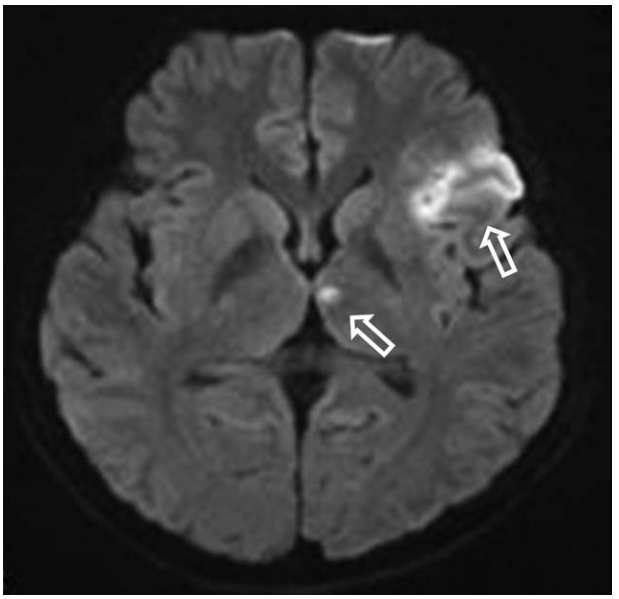

Figure 2. Brain magnetic resonance imaging was performed in the acute phase at 18 hours after the initial presentation of symptoms. Diffusion-weighted images (DWI TR/TE 4700/70) showed high signal intensity near the inferior frontal gyrus of the left frontal lobe and the left thalamus (white arrow), indicating acute infarction.

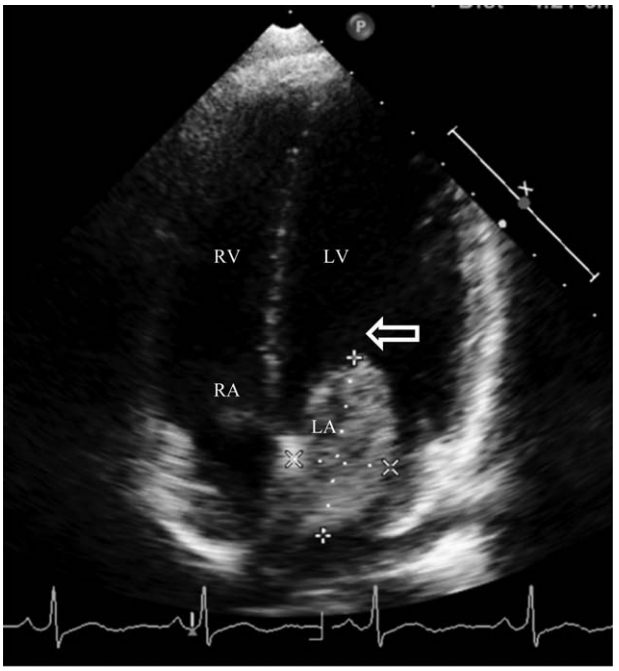

Figure 3. Echocardiography revealed a mass measuring 4.21 cm × 2.25 cm in the left atrium and attached to the interseptum. The mass was myxomatous and movable along the inflow of the mitral valve.

Her neurological examination upon discharge revealed a score of 5/5 for right extremity strength and nearly normal dysarthria. She was discharged from inpatient rehabilitation, met rehabilitation goals, and successfully regained her normal function 2 weeks after the initial symptom presentation. During the follow-up of 2 years after diagnosis, she did not present with other neurological deficits and no residual mass was observed on echocardiography. However, visual impairment of the left eye persisted.

3. Discussion

CRAO is characterized by acute, painless loss of monocular vision, and it is commonly observed in older adults.[6] No articles have reviewed cases of CRAO in the pediatric population because of its rarity, and only few reports of children and young adults with CRAO have been published.[4,6] CRAO has various etiologies, with carotid artery atherosclerosis being the most common cause in older adults.[3] On the contrary, cardiogenic embolism is the most probable etiology in children and young adults.[4,6] According to Sharma et al,[5] approximately 60% the patients <45 years of age who were diagnosed with retinal artery occlusion showed abnormal echocardiography findings. Typically, CRAO presents with other symptoms. In a study by Greven et al,[4] 20 of the 25 patients presented with accompanying symptoms observed in young patients with retinal artery occlusion. Once CRAO occurs, the function of the damaged eye cannot be recovered, and the risk of cardiovascular and cerebrovascular events increases in such patients. In addition, even life expectancy may be reduced.[12–16]

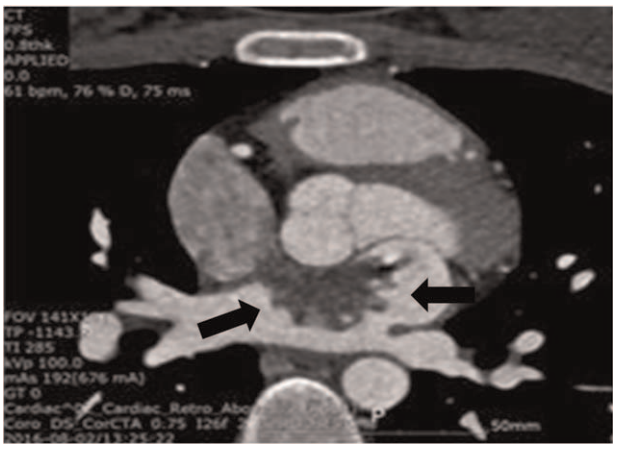

Figure 4. Cardiac computed tomography revealed an enhanced mass measuring 3 cm × 2.2 cm × 3 cm with an irregular margin on the anterior wall of the left atrium and the border of the fossa ovalis.

CRAO can be caused by cardiac myxoma, particularly in the left atrium. Myxoma may cause embolism because of its friable nature, primarily affecting the middle cerebral or supraclinoid internal carotid artery.[8,9] According to Pinede et al,[7] the location of embolism was the cerebrum in 21%, peripheral (limb) vessels in 13%, and ≥ sites in 7% of the cases, indicating that mild symptoms (red spots on the limbs) may occur without serious neurological deficit (cerebral infarction). Other than embolism, obstructive and constitutional symptoms may occur in left-sided cardiac myxoma. However, as in the case of embolism, the extent of symptoms varies. Approximately 67% of the individuals with such a condition present with obstructive signs, ranging from malaise or syncope to cardiac failure (14%–43%). Reportedly, constitutional signs account for 34% of the signs, ranging from fever to decreased general condition.[7]

Likewise, Fuchs et al[17] have assessed 17 pediatric patients with acute ischemic stroke caused by cardiac myxoma and showed that 16 patients presented with cerebral infarction with concomitant symptoms, such as obstructive symptoms, constitutional symptoms, and embolisms in the other sites.

Considering the poor prognosis of CRAO, cardiac myxoma must be diagnosed as early as possible before the development of CRAO. However, no published study has assessed CRAO caused by cardiac myxoma in pediatric patients. Therefore, in this study, we aimed to retrieve and review cases of CRAO caused by cardiac myxoma in pediatric populations.

To date, only 7 pediatric cases of CRAO caused by cardiac myxoma have been reported (1)[9,18–23] in addition to the present case. In all cases, cardiac myxoma originated in the left atrium. Such myxoma is more likely to cause CRAO as fragments from the myxoma in the left atrium can travel to the systemic vessels, such as the retinal artery, whereas those from the right atrium cannot.[24] In addition, all patients presented with other embolic symptoms, such as cerebral infarction, red spots on the limbs, and pain in the extremities, apart from visual disturbance, indicating that other embolic symptoms may appear before the occurrence of CRAO. Unlike in this case, not all patients present with obstructive or constitutional symptoms. Among other studies, only 3 of the 7 cases presented with obstructive symptoms, whereas 4 presented with constitutional symptoms. These reports indicate that CRAO is unlikely to be the initial symptom of cardiac myxoma, and similar tendency has been identified in previous studies conducted in all age groups.[25] However, unlike in this case, only 3 patients exhibited abnormal chest examination findings, whereas only 1 patient exhibited abnormal EKG findings, suggesting that a diagnostic clue must be obtained

Table 1

Review of the clinical symptoms of cardiac myxoma with central retinal artery occlusion in the pediatric population.

Reference	Age/ sex	Location of tumor	Obstructive symptoms	Embolic symptoms	Constitutional symptoms	Cardiac murmur	EKG findings	Brain imaging	Outcome after surgical removal
[18]	10/F	Left atrium, left ventricle	None	Right hemiparesis, left blindness	None	No	Normal	CT: lesions in the left internal capsule/corpus callosum, fusiform aneurysms in the left ACA, MCA	Improvement of right hemiparesis, persistent left-eye blindness, recurrence of tumor
[19]	8/M	Left atrium	None	Right hemiparesis, seizure, aphasia, red spot on the hands/ feet, left blindness	None	No	Not done	MRI: multiple cerebral lesions	Right hemiparesis, persistent visual impairment
[9]	11/F	Left atrium	None	Right hemiparesis, expressive aphasia, red spots on the hands/feet, left eye blindness	Nausea, abdominal pain	NO	Not mentioned	CT: right proximal MCA occlusion MRI: infarcts of both hemispheres MRA: normal	Persistent left hemiparesis, improvement of expressive aphasia
[20]	13/M	Left atrium	Exertional dyspnea, chest pain, syncope, loss of consciousness	Left hemiparesis, aphasia, red spots on the foot, reduced visual acuity in the right eye	Diaphoresis	Grade 3/4 pansystolic murmur over the apex with radiation into the left sternal border/left axilla	Incomplete right bundle branch block, left atrial enlargement		Visual impairment
[21]	17/M	Left atrium	Malaise	Right hemiplegia, aphasia, left blindness, pain in the legs, red spots on the hand	Night sweats, anorexia,	Not mentioned	Not mentioned		
[22]	13/F	Left atrium	Syncope	Left hemiplegia, blindness, pain in both legs, splinter hemorrhages in the nailbeds, adrenal gland	Low-grade fever	Diastolic murmur in the left fifth intercostal space and a diastolic gallop at the base	Not mentioned	Not mentioned	Death
[23]	17/M	Left atrium	Chest pain	Red spots on the hands, one-side blindness, pain in the legs	None	Diminished left carotid pulse, diminished left superficial temporal pulse	Not mentioned	Not mentioned	Not mentioned
Case 1	17/F	Left atrium	Syncope	Right side weakness/ numbness, left blindness, red spots on the foot	None	Normal	Normal	MRI: multifocal acute infarction in the left frontal lobe/left thalamus MRA: normal	Improvement of right hemiparesis, persistent left visual impairment

ACA = anterior cerebral artery, CT = computed tomography, EKG = electrocardiography, MCA = middle cerebral artery, MRI = magnetic resonance imaging.

through a thorough review of history of mild symptoms, such as vomiting, syncope, and skin rash, and that it must be determined whether additional tests, such as echocardiography, are necessary. Records of prognosis were available for only 5 of the 7 cases and most showed poor outcomes; all patients presented with persistent visual impairment, 2 presented with persistent hemiparesis, and 1 died.

In all cases including our patient, cardiac myxomas were found only after permanent blindness had occurred, but blindness was not the only symptom. In other words, we should perform detailed history and physical examination of mild symptoms, such as repeated syncope or other embolic symptoms, so that we can early detect the tumor and prevent blindness or other worst cases.

4. Conclusion

CRAO is a rare disease. Nonetheless, such a condition must be prevented to avoid poor prognosis. Other mild symptoms caused by cardiac myxoma may precede CRAO, which may be easily neglected. Therefore, we recommend the examination of nonspecific but helpful diagnostic symptoms, such as frequent syncope, recurrent cutaneous emboli, or transient ischemic attacks, and subsequent echocardiography before the occurrence of CRAO.

5. Ethical review and patient consent

The Institutional Review Board of Chonbuk National University Hospital stated that it was not necessary to achieve IRB approval for this case report, but that patient consent was required as the study dealt only with retrospective use of the patient's medical records and related images. Written informed consent was obtained from the patient before the publication of this case report and accompanying images. This study was performed by the approval of the Institutional Review Board of Chonbuk National University Research Council (CUH 2019-04-12).

Author contributions

Conceptualization: Seo Yeon Yi, Min Jeong Han, Young Hwa Kong, Chan Uhng Joo, Sun Jun Kim.
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Writing – review & editing: Young Hwa Kong, Sun Jun Kim.

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