

[ CASE REPORT ]

## Paradoxical Cerebellar Embolism Associated with Platypnea-orthodeoxia Syndrome

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### Abstract:

Platypnea-orthodeoxia syndrome is a rare clinical entity characterized by dyspnea and arterial blood deoxygenation in a sitting position. An 89-year-old woman was diagnosed with subacute cerebellar infarction. Her blood oxygen saturation decreased to 88% in a sitting position, resulting in dyspnea. Cardiological thoracic computed tomography revealed an unruptured aortic aneurysm, an enlarged ascending aorta, right atrial compression, and counterclockwise rotation of the heart. An anatomical distortion of the atrial septum induced by these abnormalities directed the atrial venous inflow such that the right-left shunt flow was exacerbated in a sitting position.

**Key words:** platypnea-orthodeoxia syndrome, paradoxical cerebellar embolism

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### Introduction

Platypnea-orthodeoxia syndrome (POS) is a rare clinical entity characterized by dyspnea and arterial blood deoxygenation in an upright or sitting position. This can occur due to the mixing of deoxygenated venous blood with oxygenated arterial blood via a shunt. Patients with an intracardiac shunt also demonstrate a secondary anatomic or functional defect. The most commonly described cause of an intracardiac shunt is patent foramen ovale (PFO), and the second-most common is either an atrial septal defect (ASD) or an atrial septal aneurysm. Although many reports have described cases of POS in which a main symptom was dyspnea, POS has rarely been associated with paradoxical embolism (1).

We herein report a case of cardioembolic cerebellar infarction with POS induced by right atrial compression and counterclockwise rotation of the heart with distortion of the atrial septum.

### Case Report

An 89-year-old right-handed woman who had exhibited a

progressive drowsy state for 12 days was brought to our hospital by ambulance. She had been initially diagnosed with chronic heart failure at her local clinic.

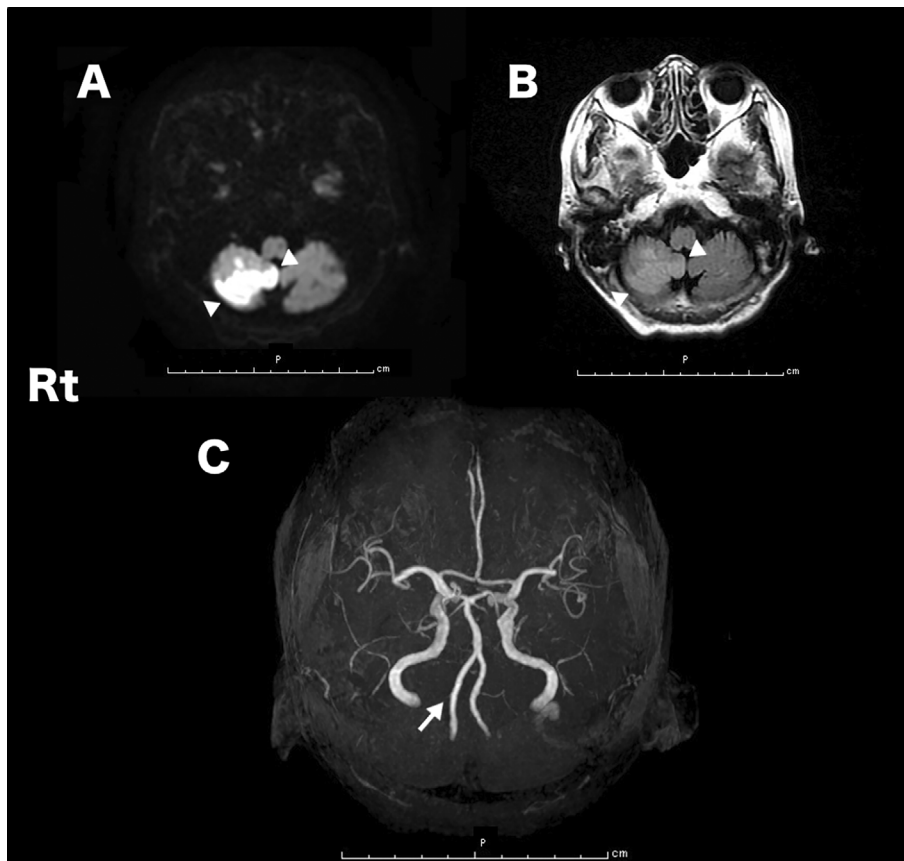
On admission, her blood oxygen saturation was 99% in a recumbent position with no abnormality of her electrocardiogram. Her drowsy state seemed to be caused by general fatigue. Neurological examinations on admission revealed normal consciousness, no eye movement disorder, no nystagmus, no dysarthria, mild left-sided weakness (positive pronator drift test and Mingazzini leg test), and severe ataxia of the right upper and lower extremities. Tendon reflexes were normal in all extremities; however, the Babinski and Chaddock reflexes were elicited on the left side. The patient's National Institutes of Health Stroke Scale score was 4 points.

Laboratory tests revealed elevated concentrations of D-dimers (16.1 µg/mL), brain natriuretic peptide (104.3 pg/mL), blood urea nitrogen (33 mg/dL), creatinine (0.91 mg/dL), and C-reactive protein (0.77 mg/dL). Magnetic resonance imaging (MRI) showed disseminated high-intensity areas in the right cerebellar hemisphere and tonsil on diffusion-weighted and fluid-attenuated inversion recovery images with no stenosis of the vertebral arteries on magnetic resonance angiography. No asymptomatic cerebral infarct

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**Figure 1.** (A) Diffusion-weighted magnetic resonance imaging, (B) fluid-attenuated inversion recovery imaging, and (C) magnetic resonance angiography in the axial view. A large subacute infarction is seen in the right cerebellar cortex (A, white arrowheads). The right posterior inferior cerebellar artery is occluded (C, white arrow).

was present in the supratentorial lesion. The right posterior inferior cerebellar artery could not be identified on magnetic resonance angiography (Fig. 1). Continuous electrocardiogram monitoring detected no atrial fibrillation.

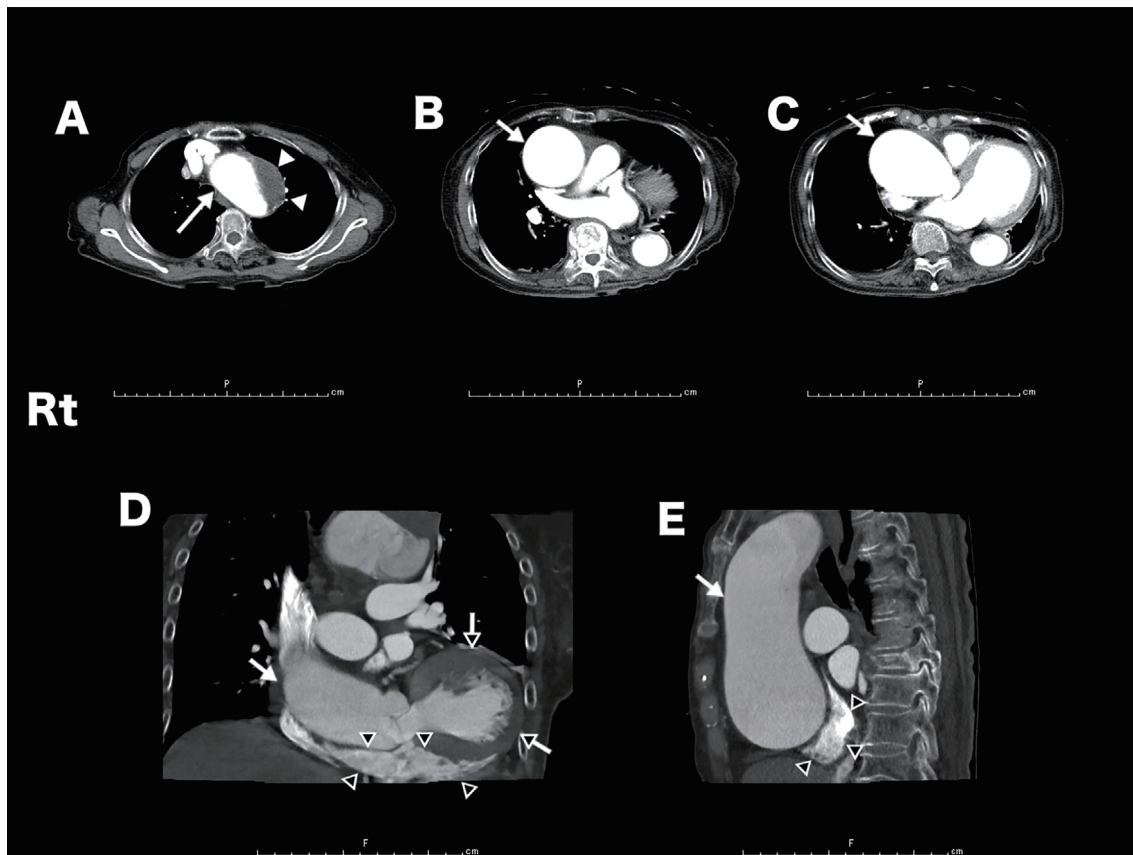
After admission, the patient's blood oxygen saturation was 95% when in a recumbent position and decreased to 88% in a sitting position, resulting in dyspnea. Enhanced thoracic computed tomography (CT) revealed a 45-mm unruptured aortic aneurysm, an enlarged ascending aorta without a mural thrombus, right atrial compression, and counterclockwise rotation of the heart (Fig. 2). Enhanced chest CT did not demonstrate the presence of pulmonary embolisms. Although no microbubbles were found in the left atrium or ventricle in a recumbent position by transthoracic echocardiography, microbubbles were observed from the right to left atrium and ventricle in the first three beats in a sitting position (Fig. 3). The Valsalva maneuver and transesophageal echocardiography could not be performed because of the patient's severe exhaustion. Ultrasonography revealed swelling of the venous wall from the superficial femoral vein to the popliteal vein without obvious venous thrombosis. Therefore, we diagnosed the patient with paradoxical cerebral embolism associated with POS.

Anatomic dilatation and distortion of the proximal ascending aorta led to right atrial compression and counter-

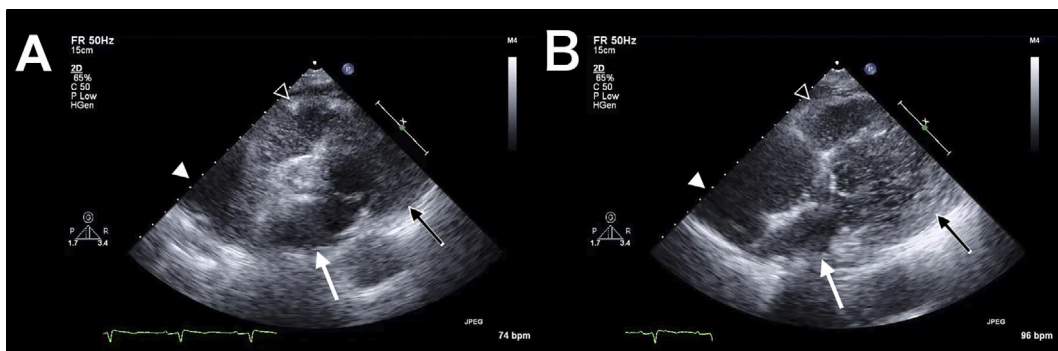
clockwise rotation of the heart, distorting the atrial septum. This anatomical distortion of the atrial septum directed the atrial venous inflow and caused narrowing of the tricuspid valve flow tract, such that the right-left shunt flow was exacerbated in a sitting position. She was treated with the intravenous administration of heparin sodium and edaravone for six days after admission and with apixaban thereafter. Her hypoxia was treated with low-flow oxygenation in a recumbent position. Her family rejected surgical treatment of the unruptured aortic aneurysm because of her advanced age, and she was transferred to an extended-care hospital.

## Discussion

POS is a relatively rare clinical disorder. Its primary mechanisms can be broadly classified as intracardiac abnormalities, extracardiac abnormalities, and miscellaneous etiologies (1). Intracardiac communication between the two atria is the most common cause of POS. Although an intracardiac shunt leading to POS has been reported in patients with ASD and PFO, PFO is the most commonly reported site of an intracardiac shunt (2). Most patients with an intracardiac shunt also demonstrate a secondary anatomic or functional defect. The most common secondary factor is an anatomic modification (dilatation and/or distortion) of the



**Figure 2.** Enhanced thoracic computed tomography in the (A-C) axial view, (D) coronal view, and (E) sagittal view. Cardiological imaging reveals a 45-mm unruptured aortic aneurysm (A, white arrowheads) and an enlarged ascending aorta (B-E, white arrows). Anatomic dilatation and distortion of the proximal ascending aorta induced by the aneurysm led to right atrial compression (D and E, black arrowheads) and counterclockwise rotation of the heart with distortion of the atrial septum (D, black arrows).



**Figure 3.** Transthoracic echocardiography in the (A) recumbent position and (B) seated position. The images depict the right atrium (white arrowhead), right ventricle (black arrowhead), left atrium (white arrow), and left ventricle (black arrow). (A) Although no microbubbles are observed in the left atrium or ventricle in the recumbent position, (B) many can be seen in the seated position.

proximal ascending aorta induced by an aneurysm, which leads to right atrial compression and atrial septum distortion, especially in a sitting position (3-5). The change in the axis of the atrial septum might also be exacerbated with age (6). This anatomical distortion of the atrial septum directs the atrial venous inflow through the ASD or PFO and causes narrowing of the tricuspid valve flow tract, resulting in exac-

erbation of the right-left shunt flow (7).

To our knowledge, the present case is the first paradoxical cerebellar embolism associated with POS to be reported, although we could not prove the presence of PFO. The patient showed mild left hemiparesis with positive Babinski and Chaddock reflexes and ataxia of the right side. Although we found no acute infarction in the right medulla on MRI, an

area of hypoperfusion might have been present.

Two reports have described paradoxical cerebral embolism induced by POS. Takashima et al. (8) reported a patient with cerebral infarction who was diagnosed with POS caused by PFO. The enlargement of the ascending aorta compressing the right atrium in the sitting position induced the atrial venous inflow through the PFO, such that the right-left shunt flow was exacerbated. In addition, Yoshida et al. (9) described a man with POS induced by the compressed right atrium secondary to an elongated aorta through the ASD with a cerebral infarction.

Cryptogenic cerebral embolism with POS is quite a rare condition, and achieving a correct diagnosis through routine examinations is difficult. It is important to consider paradoxical intracranial embolism in patients who develop dyspnea in a sitting position.

**The authors state that they have no Conflict of Interest (COI).**

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