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Case Report

# Persistent double dorsal aorta in patient with cavernous sinus thrombosis: A case report ☆

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ABSTRACT

Persistent double dorsal aorta is a rare congenital anomaly of the descending aorta in which the descending aorta is divided into 2 lumens below the ligamentum arteriosum. There are only a few cases reported to date. A 52-year-old female presented with right ptosis since 2 months prior. Neurological examination was significant for cavernous sinus syndrome in the right eye. Digital subtraction angiography revealed right cavernous sinus thrombosis and an incidental finding of double lumen descending aorta, with separation of the second lumen at the level of the thoracic aorta. Computed tomography angiography confirmed a type 2 persistent double dorsal aorta. Persistent double dorsal aorta consists of 2 types. Type 1 is the complete separation of the 2 descending aorta and type 2 is the double lumen descending aorta separated by a dividing septum. Multiplanar 3D reconstruction Computed tomography angiography or magnetic resonance angiography is important to differentiate between this anomaly and acquired conditions such as aortic dissection. In persistent double dorsal aorta, both lumens constitute the true lumen, and branch into the visceral arteries before ending up as the right and left common iliac arteries, respectively, while in aortic dissection, one is a false lumen and does not give a branch to visceral vessels. Persistent double dorsal aorta is a rare congenital anomaly of descending aorta which manifests as 2 separate aorta or 2 lumens of aorta separated by a dividing septum. Knowledge of this anomaly is paramount for interventional neuroradiologists to distinguish it from acquired lesions.



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## Introduction

Persistent double dorsal aorta is a rare congenital anomaly of the descending aorta in which the descending aorta is divided into 2 lumens below the ligamentum arteriosum. There are only a few reported cases of this condition. A systematic review in 2017 found eleven cases of persistent double dorsal aorta [1]. More recently, an additional case of persistent double dorsal aorta in neonates was reported [2]. Three other cases described similar anomalies of the abdominal aorta, termed double abdominal aorta [3–5].

In this case, we report a case of persistent double dorsal aorta which was found incidentally, in a patient who underwent angiography for another diagnosis. The patient's descending aorta consists of 2 lumens, separated by a septum, from the thoracic aorta up to the aortic bifurcation. Each lumen terminated as the right and left common iliac arteries. We also review the previous cases of persistent double dorsal aorta and the implications regarding this anomaly, which may provide additional knowledge for interventional neuroradiologists.

## Case report

A 52-year-old female presented with a chief complaint of right eyelid drooping, which worsened gradually since 2 months prior. She also had a right-sided headache and double vision, which were exacerbated by right conjugate eye movement. Her symptoms worsened over time until she decided to seek medical treatment. She had a history of hypertension, gallstones, and fatty liver, but no history of head trauma, chest trauma, or any sign and symptoms of infection.

Her neurological examination was significant for right-sided cavernous sinus syndrome. No bruit was heard over her eyelid. A non-contrast head computed tomography (CT) scan did not show a clear pathological process nor any sign of paranasal infection, and the initial working diagnosis was an indirect carotid-cavernous fistula. A digital subtraction angiography was ordered to confirm the diagnosis. When navigating the descending aorta, we noted the double lumen of the descending aorta, with the separation of the second lumen starting from the level of the thoracic descending aorta (Fig. 1). CT angiography performed later revealed a congenital anomaly of persisted double descending aorta (Fig. 2).

The patient was eventually diagnosed with right-sided cavernous sinus thrombosis and received fondaparinux subcutaneously for 5 days, followed by warfarin. Her symptoms gradually improved in 1 month. As for the persistent double dorsal aorta, she did not have any complaints of chest or abdominal pain or any symptoms related to the anomaly up until 6 months follow-up.

## Discussion

Anomaly of the descending aorta is extremely rare. In this case, we found a case of persistent double dorsal aorta with 2 lumens of descending aorta separated by a septum. The septum started to divide the aorta from the thoracic descending aorta to the aortic bifurcation. The celiac artery arises from the right lumen (Fig. 2D), and the renal artery arises from the corresponding ipsilateral lumen (Fig. 2E). Each lumen terminates as the right and left common iliac artery (Fig. 2F). We didn't find any other congenital anomalies in the heart or other great vessels in this case.

Previous case reports found 2 variants of persistent double dorsal aorta. Type 1 is a complete separation of the 2 dorsal aortas and type 2 is a double lumen of the descending aorta separated by a dividing septum [1,6,7]. The paired branches of the descending aorta, such as the intercostal, lumbal, and renal arteries both arise from 1 aorta in the first type, but arise separately on each side of the corresponding aorta in the second type. The unpaired branch arises from 1 aorta in both types.

Our case was a type 2 persistent double dorsal aorta, which is less common than type 1. There are only 2 previous cases, which were reported by Trubnikov and Hristova [8,9]. The first case by Trubnikov was associated with an anomaly of the vessel in which a branch of the right lumen crossed the left lumen into the left renal artery to supply the upper portion of the left kidney [8]. The second case by Hristova was more similar to our case, with a double lumen aorta separated by a septum from the level of the fourth thoracic vertebra to aortic bifurcation and without other anomalies of the heart or great vessel [9].

In contrast to type 2, type 1 is more commonly associated with other congenital anomalies. Formanek et al. [7] reported a case of double dorsal aorta in a 2-years-old boy associated with complex congenital heart disease, including dextrocardia, endocardial cushion defect with a common atrioventricular valve, double outlet venous ventricle with the aorta anterior to the pulmonary artery, pulmonary stenosis, bilateral trilobate lung, and asplenia. Mills and Gets [1] reported other cases of type 1 double dorsal aorta associated with bilateral bilobate lung, bilateral tendinous attachment of the diaphragm to the Thoracic-10 vertebral body, apparent suprarenal tissue cranial to a diaphragm, and flattened liver. Other congenital anomalies reported were renal agenesis, congenital kyphosis, vaginal agenesis, and horseshoe adrenal gland [1]. Only the case reported by Karadeli et al., [6] in 2009 did not report any congenital anomaly associated with type 1 persistent double dorsal aorta.

Differentiating between persistent double dorsal aorta and other acquired conditions, such as aortic dissection, can be challenging. The differentiation can be made based on clinical and imaging findings. Type 2 persistent double dorsal aorta is usually asymptomatic, in contrast to aortic dissection which usually has more prominent clinical manifestations such as severe chest pain or cardiogenic shock. Physical examination in aortic dissection also discloses abnormal findings like diastolic murmur, pulsus paradoxus, hypovolemic shock, or paraplegia [10]. Nowadays, noninvasive imaging such as a CT scan or magnetic resonance imaging (MRI) has replaced invasive imaging in the diagnosis of aortic disease. Multiplanar 3D reconstruction of CT or MR angiography is a useful tool to disclose the anatomy of both lumens and their relationship with other visceral vessels. In type 2 persistent double dorsal aorta,

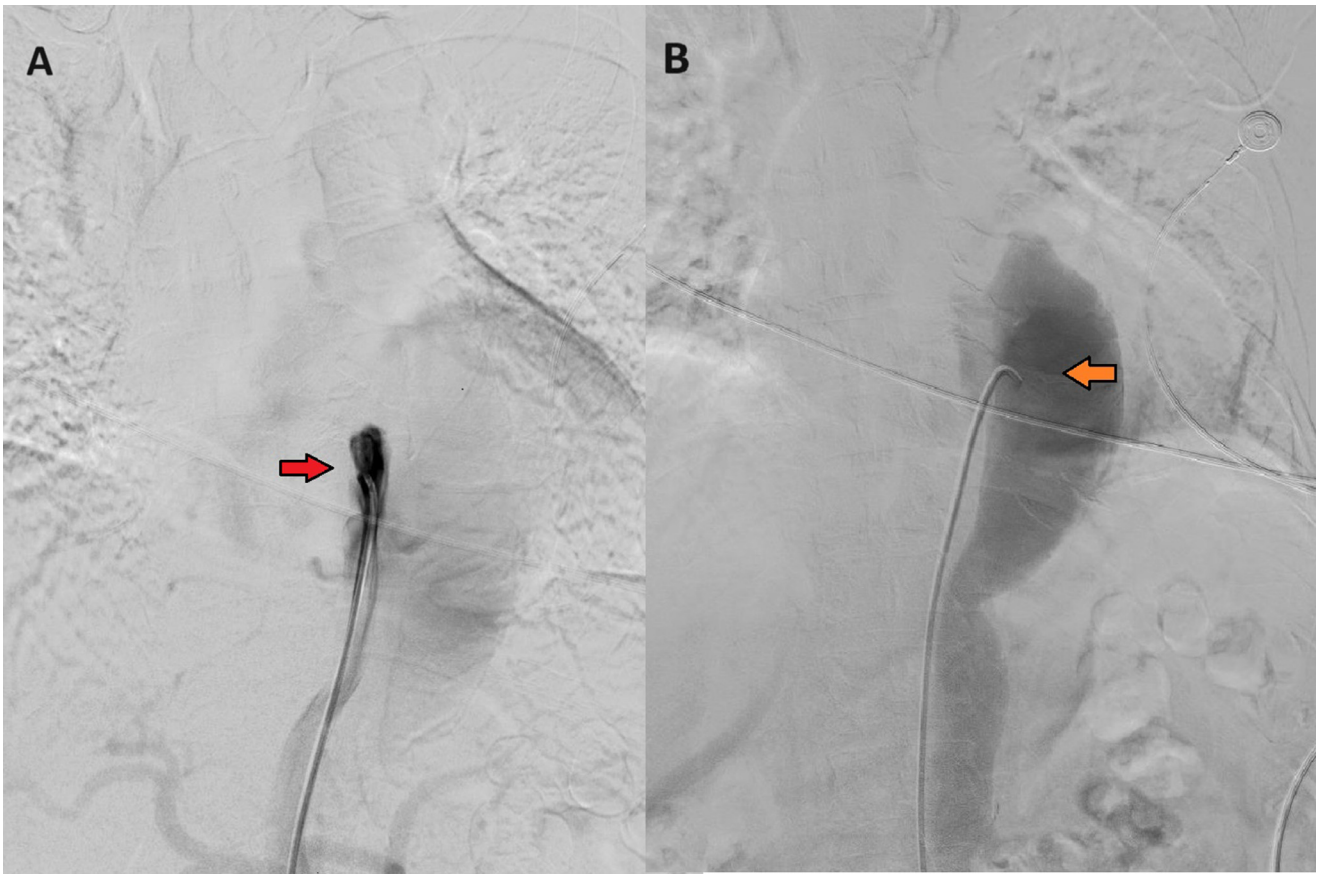


**Fig. 1 – Thoracic aorta aortogram revealed a double lumen of the aorta. (A) The tip of the catheter is at the first lumen (red arrow), which was connected to the right femoral artery where the puncture site was located. (B) The tip of the diagnostic catheter entered the second lumen (orange arrow) at the level of the thoracic aorta. Note that the contrast filled the second lumen on the left side of the first lumen.**

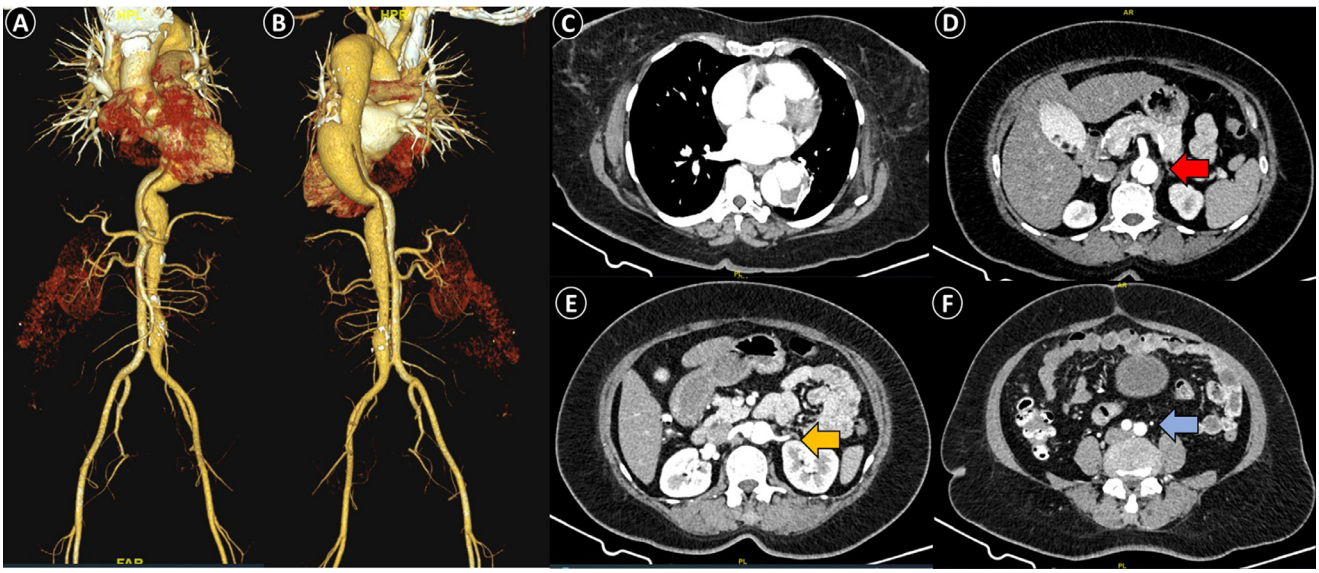


**Fig. 2 – Three-dimensional reconstruction (3D) computed tomography angiography (CTA) anterior (A), and posterior (B) views revealed persistent double dorsal aorta. The axial view shows the celiac artery arises from the right lumen (red arrow) (D), the right renal artery from the right lumen, the left renal artery from the left lumen (orange arrow) (E), and each lumen terminates as right and left common iliac artery (blue arrow) (F).**

both lumens are true lumens, and each lumen gives a branch to the visceral vessel and terminates as the right and left common iliac artery respectively. In aortic dissection, one lumen is the true lumen and the other is a false lumen [11]. The true lumen gives branches to visceral vessels and terminates at the aortic bifurcation as both the right and left common iliac arteries, while the false lumen does not give branches.

## Conclusion

Persistent double dorsal aorta is a rare congenital anomaly of the descending aorta that manifests as 2 separate aortas or 2 lumens of the aorta separated by a dividing septum. Knowledge of this anomaly is paramount for interventional neuroradiology to distinguish it from acquired lesions. In persistent double dorsal aorta, both lumens are true lumens, and each lumen gives a branch to the visceral vessel.

## Patient consent

The subject described in the case report has provided consent for publication.

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