Left Ventricular Noncompaction Cardiomyopathy as a Potential Cause of Bilateral Posterior Cerebral Artery Stroke – a Rare and Unique Clinical Occurrence

Marwa Elnazeir
Lisa Nobel
Siddharth Narayanan
Fahed Darmoch
Francesco Massari
Timothy Fitzgibbons
Raphael A. Carandang
Adalia Jun-O’Connell

Corresponding Author: Marwa Elnazeir, e-mail: marwa.elnazeir@umassmemorial.org

Conflict of interest: None declared

Patient: Male, 63-year-old
Final Diagnosis: Occurrence of bilateral PCA infarcts with LVNC cardiomyopathy
Symptoms: Acute vision loss in both eyes • dysarthria
Medication: —
Clinical Procedure: Emergency endovascular thrombectomy
Specialty: Neurology

Objective: Rare co-existence of disease or pathology

Background: Bilateral posterior cerebral artery (PCA) occlusions are exceedingly rare, and are considered a devastating phenomenon that presents as cortical blindness. Predominant causes of PCA infarcts include cardiac and arterial embolisms. Left ventricular noncompaction (LVNC) cardiomyopathy is also an extremely rare cardiopathology. Several reports describe stroke as a potential manifestation of LVNC, but bilateral PCA infarcts are likely also caused by underlying LVNC cardiomyopathy, although this has not yet been reported.

Case Report: A 63-year-old man presented to the emergency department of an outside hospital with acute vision loss in both eyes and dysarthria. His neurological examination necessitated an emergent stroke evaluation. His electrocardiogram and telemetry at admission did not reveal arrhythmia. He underwent an emergency endovascular thrombectomy at our facility. During the post-intervention stroke workup, a transthoracic echocardiogram with contrast showed left ventricle dilation, with an ejection fraction (EF) of 29%. Subsequent cardiac magnetic resonance imaging confirmed the presence of LVNC cardiomyopathy. He was started on therapeutic anticoagulation (apixaban) and remained stable neurologically during the 3-month followup, with some residual visual field deficits. His cardiac outcome also improved (stress test was unremarkable for any cardiac ischemia, and an echocardiogram showing improved EF of 40%).

Conclusions: Our report is distinct, as it presents 2 exceedingly rare events in a patient: the occurrence of simultaneous bilateral PCA infarcts and LVNC cardiomyopathy. Prompt and accurate diagnosis was pivotal to the successful management of both conditions. Prospective studies are warranted to further knowledge of LVNC pathophysiology and the occurrence of stroke in such patients so that comprehensive management plans can be devised.

Keywords: Cardiomyopathies • Infarction, Posterior Cerebral Artery • Stroke

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/931103
Background

Stroke, either hemorrhagic or ischemic in cause, is the second leading cause of death and the third most common worldwide cause of disability [1]. In ischemic strokes, the middle cerebral artery is the most commonly affected branch of occlusion, whereas the incidence of infarction in the posterior cerebral artery (PCA) territory or in its branches is low, with an incidence of 5-10% in a general stroke population [2,3]. Specifically, the incidence of bilateral PCA occlusions is exceedingly rare and considered a devastating phenomenon presenting as cortical blindness [2]. Cardiac or arterial embolisms are thought to be the predominant causes of PCA infarcts [2].

Left ventricular noncompaction (LVNC) is a rare cardiomyopathy characterized by prominent trabeculae and deep intertrabecular recesses that communicate with the left ventricular cavity and a thin and compacted epicardial layer [4]. LVNC has an incidence of 0.05% in adults [5], and men are more commonly affected than women [6]. It has an ill-defined natural history and clinical manifestation [7], although it is suggested to result from an intrauterine arrest of the left ventricular myocardium compaction process [8]. LVNC cardiomyopathy has been associated with ischemic strokes [9]. However, whether or not LVNC is a likely cause of bilateral PCA infarcts, is not known.

We present a very unique case of a patient who suffered simultaneous acute ischemic strokes on the bilateral PCA distribution, very likely due to an LVNC cardiomyopathy that was identified during the stroke workup, and after excluding other causes. He underwent a successful and uncomplicated superselective microcatheter intra-arterial alteplase (t-PA) administration for the bilateral PCA occlusions. Rapid and timely diagnosis was pivotal to the successful management of both conditions.

Case Report

A 63-year-old man arrived at the emergency department (ED) of an outside hospital with acute vision loss in both eyes and slurred speech. He had a past medical history of chronic smoking, hypertension, chronic back pain, acid reflux, and seasonal allergy. His home medications included amlodipine (10 mg daily per os), cyclobenzaprine (10 mg, per os, 3 times daily as needed for muscle spasm), flucasicone nasal spray (as needed for seasonal allergy), naproxen (375 mg per os every 8 h as needed for chronic back pain), and omeprazole (40 mg per os daily). He reached the ED within 15 min of his acute symptom onset. These symptoms were never experienced before, were sudden in onset, and with no pain associated with the vision loss. Clinically, he did not show any signs of acute heart failure (HF) that were exacerbated upon presentation, including any lack of shortness of breath or pitting edema.

While he was awake and alert upon arrival to the ED, he was hypertensive, with a systolic blood pressure of 190-200 mmHg. His random blood glucose level was 102 mg/dL. Urgent neurological assessment, measured by the National Institute of Health Stroke Scale (NIHSS) was 4 (3 for total blindness, and 1 for speech abnormality). This necessitated an emergent code stroke activation. His electrocardiogram showed a normal sinus rhythm and subsequent telemetry monitoring did not reveal any arrhythmia. An emergent computerized tomography (CT) of the head showed no evidence of intracranial hemorrhage, while a CT angiogram of the head and neck showed bilateral right P2 and left P3 PCA focal occlusions, respectively (Figure 1), with no evidence of atherosclerosis or thrombosis within the basilar artery.

The patient received an intravenous t-PA (alteplase 0.9 mg/kg), and was transferred to our comprehensive stroke center. Because of the distal location of the occlusions and the presence of systemic t-PA therapy on-board, the patient underwent a successful and uncomplicated superselective pharmacological thrombectomy treatment via microcatheter, delivering intra-arterial t-PA infusion (3 mg each for the left and right P2 segments). This resulted in recanalization of the right P2 branches, and improved flow within the left P2 distal branches. The successful reperfusion of the bilateral PCA occlusions caused significant improvement of the antegrade flow within the corresponding PCA vascular territories (Figure 2).

As part of the stroke workup, the telemetry during hospitalization did not show any evidence of atrial fibrillation (AF) or any other arrhythmia. The subsequent 30-day cardiac monitor also did not identify any cardiac arrhythmia. The thrombophilia panel (or hypercoagulable panel) was also unremarkable. Magnetic resonance imaging (MRI) of the brain post-intervention demonstrated the non-salvageable areas of ischemia in the bilateral PCA vascular distribution (Figure 3).

A transthoracic echocardiogram (TTE) with contrast revealed a dilated left ventricle (LV), and a severely decreased systolic function with an EF of 29% with a concern for LVNC (Figure 4A, 4B). The patient’s brain natriuretic peptide level was grossly abnormal (1459 pg/mL; the normal value is less than 100 pg/mL). He had elevated low-density lipoprotein levels (117 mg/dL) but his HbA1c levels, anti-nuclear antibody titer, erythrocyte sedimentation rate, and C-reactive protein levels were all within normal limits. A subsequent cardiac MRI revealed hypertrabeculation of the LV, meeting the criteria for noncompaction focally at the apex (the ratio of noncompacted to compacted myocardium was above 2.3), with no intraventricular thrombus (Figure 4C). The imaging also revealed a dilated LV, globally
Figure 1. Computerized tomography angiogram of the head and neck. The black arrows point to the (A) right P2 and (B) left P3 posterior cerebral artery focal occlusions.

Figure 2. Cerebral angiogram post-pharmacological thrombectomy. Superselective microcatheter intra-arterial administration of tissue plasminogen activator resulted in reperfusion of the bilateral posterior cerebral artery (PCA) occlusions, with significant improvement of the antegrade flow within the corresponding PCA vascular territories.

Figure 3. Magnetic resonance imaging (MRI) of the brain. A post-intervention MRI of the brain confirmed bilateral posterior cerebral artery (PCA) infarcts, very limited in extension at the level of the left PCA vascular territory.
hypokinetic, with no delayed gadolinium enhancement, with an overall LV-EF of 25%.

The patient passed a swallow (dysphagia) evaluation and was started on daily aspirin (81 mg) and atorvastatin (80 mg). After a discussion with cardiology, the decision was made to start him on therapeutic anticoagulation 7 days post-stroke to minimize the risk of hemorrhagic transformation in the infarcted areas. The patient was subsequently started on apixaban (preferred, as it has a lower risk of bleeding given the size of the stroke), and remained stable neurologically at the 3-month outcome check, with a residual left visual field cut. His cardiac outcome also showed improvement, with a subsequent stress test not revealing any cardiac ischemia, and follow-up 2-dimensional (2D) echocardiography (within 2 months) showing an improved EF of 40%, which was likely due to the initiated guideline-directed medical therapy for HF.

Discussion

Several reports describe stroke as a potential manifestation of LVNC cardiomyopathy [6,7,9-15], but LVNC cardiomyopathy as a likely cause for a PCA infarct is extremely rare [16]. This is the first report, to the best of our knowledge, that identifies LVNC cardiomyopathy as a highly suspected cause for simultaneous bilateral PCA infarcts. We consider LVNC cardiomyopathy to be a likely cause in this particular case because no other abnormality (such as AF, or hypercoagulable state) was identified. Furthermore, we speculate that his underlying medication usage of a calcium channel blocker and non-steroidal anti-inflammatory drugs (NSAIDs) did not likely contribute to his HF, given the lack of history of medication overuse, limited use of NSAIDs, and his previous tolerance to amlodipine for his chronic hypertension, without any direct complications. Our patient did not show any clinical signs or symptoms of HF at the time of admission. Therefore, after excluding cardiac arrhythmia, we strongly speculate that the presence of LVNC cardiomyopathy likely contributed to the patient’s strokes.

The occurrence of stroke in patients with LVNC cardiomyopathy is 1-2% per year with a total risk of thromboembolism of about 21-38% [17]. While PCA territory lesions caused by various stroke mechanisms are less well defined compared with those of other vascular territories, cardioembolic etiology may produce significant infarcts due to a lack of collateral flow in the acute process [18-20]. The frequency of thromboembolism is variable in adults (13-24%), with AF or systolic dysfunction known to increase the risk of cardioembolic stroke [5,21]. Two prospective studies on adult patients with LVNC cardiomyopathy report a 21-24% risk of cerebral embolism during the followup period [6], which is considered to be a higher risk than that in the non-LVNC population, considering the typical cumulative recurrent stroke risk is considered to be 3.1% at 30 days or 11.1% at 1 year [22].

We suspect that the bilateral PCA occlusions manifested in our patient as a result of a potential cardioembolic event due to LVNC cardiomyopathy. The clinical manifestations of LVNC cardiomyopathy exist on a wide spectrum, ranging from asymptomatic to the classic triad of HF, arrhythmias, or thromboembolic events [4]. Cardioembolism as a result of rare cardiomyopathies can manifest as stroke, likely as a result of atrial arrhythmias or impaired EF [5]. Our patient did exhibit a severely compromised systolic function with an EF of 29% during the stroke workup period. The LV dysfunction is most

Figure 4. A 2D echocardiogram showed severely reduced left ventricle (LV) function and prominent trabeculations consistent with left ventricular noncompaction (LVNC), and cardiac MRI revealed severely reduced LV function with focal LVNC. (A, B) 2D echocardiogram showing (A) end-diastolic apical 4-chamber view, and (B) a short axis view demonstrating reduced LV function and prominent apical trabeculations. (C) In the end-diastolic frame of the cardiac MRI, the ratio of noncompacted myocardium (NCM) to compacted myocardium (CM) was greater than 2.3 at the apex, consistent with LVNC.

Elnazer M. et al: Co-occurrence of LVNC and bilateral PCA infarcts
© Am J Case Rep, 2021; 22: e931103

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

This work is licensed under Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)
commonly reported and attributed to diminished microcirculation in the subendocardial layer of the heart [6]. A recent meta-analysis indicated that the LV-EF, and not the extent of trabeculation, was an important determinant of adverse outcomes in LVNC patients [23]. In patients with embolic events, in vivo blood clots within the noncompacted layer have been documented [24]. The simultaneous use of echocardiography, either transthoracic or transesophageal, along with cardiac MRI, can tremendously help improve the accuracy of the findings and increase the sensitivity of testing [25]. Diagnosis of LVNC cardiomyopathy is primarily based on 2D echocardiographic findings, with proposed diagnostic criteria suggested by 3 different authors [25].

Owing to a reduced EF observed in our patient, anticoagulation therapy was initiated to reduce the risk of potential thromboembolism. Anticoagulation is recommended, especially in patients with a history of thromboembolism, AF, or reduced EF [6,26]. The CHADS<sub>2</sub>/CHADS<sub>2</sub>-Vasc score may play a role in decision-making regarding oral anticoagulation for patients with LVNC cardiomyopathy [27]. Due to limited knowledge about LVNC pathophysiology, no specific management is currently recommended, with existing approaches focusing on symptom-targeted therapy [15]. Some investigators recommend long-term prophylactic anticoagulation for all patients with LVNC cardiomyopathy, regardless of whether they have experienced thromboembolic complications, and regardless of the degree of LV dysfunction. However, such a course should be approached with prudence, after carefully weighing the benefits versus risks to identify those who would most likely benefit from this strategy [11]. Patients with LVNC and HF with reduced EF are treated according to ACC/AHA guidelines for chronic HF [28]. This includes initiation of beta-blockers, ACE inhibitors/AT2 receptor antagonists, and mineralocorticoid receptor blockers (spironolactone or eplerenone). If after initiation of pharmacotherapy, HF patients still have reduced systolic function (<35%) and are symptomatic at 3 months’ time, consideration should be given to primary prevention of sudden cardiac death and cardiac resynchronization therapy [28].

Conclusions

Our report is distinct, as it presents 2 exceedingly rare events in a patient: the occurrence of simultaneous bilateral PCA infarcts and LVNC cardiomyopathy. The LVNC was identified by TTE and cardiac MRI findings during the stroke workup. Prompt and accurate diagnosis was pivotal to the successful management of both conditions. Prospective studies are warranted to further knowledge on LVNC pathophysiology and the occurrence of stroke in such patients. This will help outline more effective and comprehensive management plans for such conditions.

Conflict of Interest

None.
References: