Acute Ischemic Stroke and Undiagnosed Heart Failure in a Patient with Incidental Dandy-Walker Syndrome: A Case Report

Zainab Krayem, Faria Tazin MD, Alexis Torres DO, Jeanmarie Petersen DO, Arthur Kwok MD
AtlantiCare Regional Medical Center, Atlantic City, N.J., U.S.A.

Introduction:

Dandy-Walker syndrome (DWS) is a rare congenital condition, occurring in approximately 1 in 25,000 to 1 in 35,000 live births. Its clinical presentation is highly variable, with some individuals remaining asymptomatic, and others with complex variations.[5] Reports of stroke in patients with DWS remain limited.
This case highlights the occurrence of acute stroke and undiagnosed heart failure in a patient with an incidental finding of Dandy-Walker syndrome.

Presentation:

A 38-year-old medically noncompliant male with past medical history of hypertension, cataract (left eye) diagnosed at 27 years old, and an unspecified heart murmur presented to the ED with left-sided facial droop, voice changes, right arm weakness, and visual disturbances. Symptoms began the prior day, and he admitted to antihypertensive non-adherence. NIH Stroke Scale score was 4.

Vitals: Afebrile, HR 108, BP 164/130. Given IV labetalol and nitroglycerin.
Labs: Elevated troponin (102), proBNP (8840), BUN (32), creatinine (1.78), AST (84), ALT (72), sodium (130), and proteinuria on urinalysis.
Imaging. MRI confirmed an acute left frontal lobe infarction and cerebellar vermis atrophy, consistent with Dandy-Walker malformation. CTA head/neck showed no significant stenosis.

Neurology was consulted, permissive hypertension was allowed (SBP 180-220), and an echocardiogram was ordered. Fall and aspiration precautions were implemented, and PT/OT/speech therapy were consulted.

Hospital Course

- Day 1: ECG suggested left ventricular hypertrophy (LVH), and echocardiogram revealed an ejection fraction (EF) of 25-30% with severe global systolic dysfunction. Dual antiplatelet therapy was initiated.

- Days 2-4: Blood pressure remained elevated but improved with adjustments to antihypertensive therapy. Cardiac catheterization was postponed due to uncontrolled hypertension and worsening kidney function. A wearable cardiac defibrillator was placed.

- Day 5 Cardiac catheterization showed no significant coronary artery disease. Hematology ruled out a hypercoagulable state.

- Day 10: Patient had an episode of supraventricular tachycardia. A transesophageal echocardiogram revealed severe systolic dysfunction (EF 20-25%), stage III diastolic dysfunction, and left ventricular trabeculations concerning for noncompaction or thrombus. Anticoagulation was initiated after a repeat head CT ruled out bleeding.

- Day 13: The patient was discharged with anticoagulation and blood pressure management in addition to PCP follow up.

Final Diagnosis List:

•Acute CVA

•Acute kidney injury

•HFrEF (EF = 20-25%)

•Non-ischemic cardiomyopathy

•Hypertensive heart disease

•Dandy-Walker Syndrome

•Glaucoma

•Hyperlipidemias:

Discussion

One systematic review that studied non-neurologic comorbidities associated with DWS found that ocular abnormalities were common, with cataracts diagnosed in 20% of cases. Notably, this patient was diagnosed with a cataract in his left eye at 27 years old. The same study also reported that HF was prevalent in 9.8% of cases.
While there is no routine screening for DWS and routine EKGs are not standard of care in our patient population, this case emphasizes the important role of routine primary care visits. Regular visits could have improved this patient's medication compliance, and a comprehensive history and physical exam might have raised suspicion and prompted earlier diagnostic workup. With an earlier diagnosis of DWS, there is a possibility cardiologic effects could have been better mitigated or managed.

This case shows the importance of thoroughly evaluating stroke etiology in younger patients. The patient's congenital anomaly, may have contributed to both his cardiovascular and neurological findings. As such, early cardiovascular imaging and assessment should be considered in patients with congenital anomalies like DWS to aid in timely diagnosis and management.

Conclusion:

This case illustrates how undiagnosed heart failure may have contributed to acute ischemic stroke in a patient later diagnosed with DWS. It shows that regular primary care visits could have improved medication adherence and how thorough history, and physical exam could have prompted earlier diagnostic workup, potentially leading to earlier identification of the congenital anomaly and proper management of arising comorbidities. A multidisciplinary approach remains essential in managing patients with complex comorbidities.

References: