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EC NEUROLOGY
Mini Case Study

Ischemic Stroke in Eisenmenger Syndrome: A Challenging Case with Clinical-Imaging Discrepancy

Mojtaba Rismanchi*

Neurologist, Clinical Neurology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran

***Corresponding Author:** Mojtaba Rismanchi, Neurologist, Clinical Neurology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran.

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Abstract

Eisenmenger syndrome can be complicated with hemorrhagic and thrombotic events. Cerebral hemorrhagic and ischemic entities may be confusing in case of eisenmenger syndrome. High hemoglobin concentration produces misleading clues in cerebral computed tomography imaging. Herein a challenging cerebral computed tomography of a patient with eisenmenger syndrome is discussed in the setting of new onset focal neurologic signs that was revealed further to be stroke. It is finally proposed that clinical judgment is superior to imaging studies in central nervous system assessments.

Keywords: *Eisenmenger Syndrome (ES); Ischemic Stroke; Clinical-Imaging Discrepancy*

Introduction

Eisenmenger syndrome (ES) is caused as a result of right to left cardiac shunt. It is related with several pathological events including hypoxia and increase in hemoglobin concentration [1]. Cerebral complications of ES are stroke and cerebral abscess which are mainly due to right-left cardiac shunt [2]. The underlying pathophysiology for cerebral complications are paradoxical embolism and hyperviscosity [1]. Bleeding and thromboembolic events both occur in ES due to blood pressure and coagulation abnormalities [1,3]. It is also demonstrated that iron deficiency in ES results in microcytosis and aggravation of thrombosis [3]. All of these evidences imply that ES poses difficulty both in the diagnosis and treatment of its complications. In this article a case of ES with neurological manifestation and a hard to diagnose computed tomography (CT) imaging is reported.

Case Report

A 24 year old male with history of congenital heart disease complicated with ES was brought to the emergency department with acute onset right sided weakness and disorientation. On examination he was awake but had Brocca and mild degrees of Wernicke aphasia, had right sided weakness with muscle power of 3 out of 5 and right sided babinski sign. CT imaging of the patient was obtained (Figure 1), with the findings of dense venous structures filled with blood and dense fissures resembling blood, indicating both Cerebral Venous Thrombosis (CVT) and Subarachnoid hemorrhage (SAH). However, focal neurologic findings in examination were compatible with arterial infarct in the territories of left Middle Cerebral Artery (MCA). High hemoglobin (Hb) and hematocrit (Hct) values explain the densities in CT as the effect of high Hb concentration in cerebral vessels. Considering that the CT alone is misleading in case of ES, the patient was

treated with normal stroke treatments including Aspirin (80 mg daily) and Atorvastatin (40 mg daily). Subsequent CT scans revealed arterial infarct in the territories of left MCA as previously dictated by examination (Figure 2). Phlebotomy was instituted for the patient as a recommendation from Hematologist and search for underlying causes of stroke including endocarditis, intra-cardiac clot and vascular abnormalities and sources of emboli were performed; they were however negative.

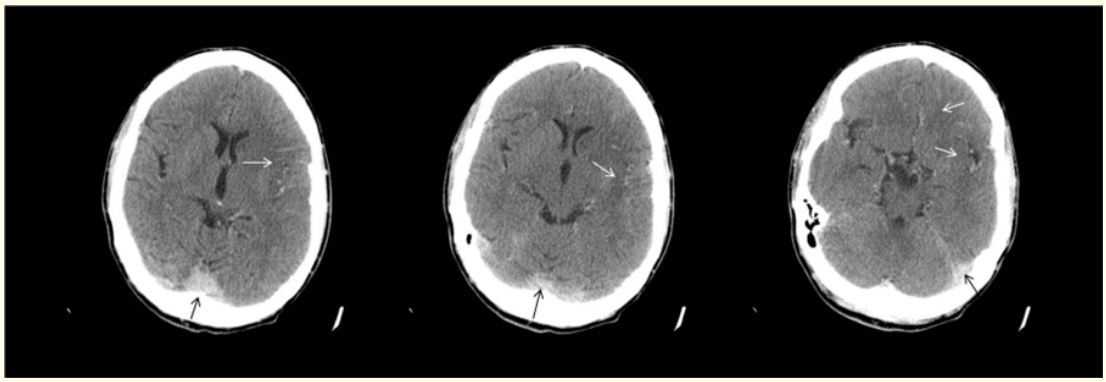

Figure 1: *Three cuts from primary axial CT imaging of the patient presented to the Emergency department with probable clues to CSVT (black arrows indicate possible cord sign) and SAH (white arrows indicate hyperdensities in fissures).*

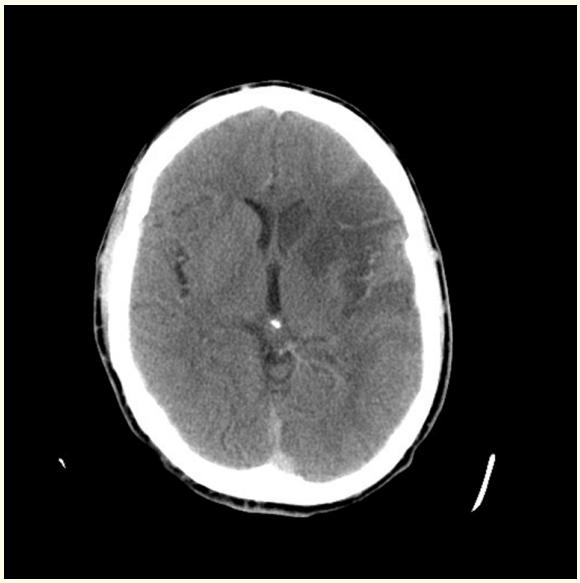

Figure 2: *Consecutive axial CT imaging shows hypodensity in the territory of left middle cerebral artery indicating infarction as the underlying cause as dictated by physical examination.*

Discussion and Conclusion

ES-patients are predisposed to thrombotic complications. Stroke, Brain abscess and predisposition to hemorrhage are possible complications in ES. Main cerebral thrombotic events in neurology are stroke and cerebral venous thrombosis and main hemorrhagic events are intracranial and subarachnoid hemorrhages.

In the present case, hyper-density of cerebral venous sinuses was noticed in CT scan that indicated CVT along with hyper-density of brain fissures resembling SAH [4,5]. Therefore, according to CT findings, SAH and CSVT were possible events. In laboratory investigations, high Hemoglobin concentrations (19 g/dl) and Hematocrit values (Htc: 46%) along with low Mean Corpuscular Volumes (MCV: 66 fL per cell) were found compatible with ES [6]. Low MCV values interpreted as iron deficiency can also be a predisposing factor for thrombotic events. Based on neurological examinations that showed neurological deficits compatible with brain arterial territories, the imaging findings were attributed to the high hemoglobin concentrations and finally he was managed as a case of young adult Cerebrovascular Accident (CVA). Furthermore, the hyper-densities in the brain fissures were, as contrary to SAH, along the course of arterial path resembling source images of enhanced CT angiography.

The key element of neurology is localization and then hypothesizing on the pathophysiology of events according to onset and course of presentation which finally yields the diagnosis. Supposing that a new lesion has happened in eloquent brain areas, as in the present case, findings on physical examination are superior to imaging findings. Indeed, those imaging findings compatible with examinations are pertinent to the present neurologic derangement. In other words, in neurology and neurosurgery, imaging is not diagnosed nor is treated rather it can only verify the clinical diagnosis.

Recommendations

In ES phlebotomy is used to decrease the viscosity; however excessive phlebotomy can contribute to iron deficiency and microcytosis and as a result its excessive use can exacerbate viscosity and thromboembolic events [7, 8]. The treatment of ES with stroke is critical in defining whether thrombotic mechanism, embolic mechanism or both are responsible. A thorough search for embolic foci should be considered, these include search for thrombosis in lower extremity veins and thrombosis hidden in pelvic veins. Finally regarding thrombosis and vascular events in polycythemia, it is reported in case of Polycythemia Vera that maintaining Hct<45.5% has been associated with better cerebral perfusion, lower whole blood viscosity and Hct<45% with higher survival rates and lower thrombotic events [9].

Conflict of Interest

The author declares that there is no conflict of interest related to the present study and its publication.

Bibliography

1. Beghetti M and Galiè N. "Eisenmenger Syndrome. A Clinical Perspective in a New Therapeutic Era of Pulmonary Arterial Hypertension 53.9 (2009): 733-740.

2. Kaemmerer H., *et al*. "The Adult Patient with Eisenmenger Syndrome: A Medical Update After Dana Point Part I: Epidemiology, Clinical Aspects and Diagnostic Options". *Current Cardiology Reviews* 6.4 (2010): 343-355.

3. Trojnarska O and Plaskota K. "Therapeutic methods used in patients with Eisenmenger syndrome". *Cardiology Journal* 16.6 (2009): 500-506.

4. Edlow JA and Caplan LR. "Avoiding pitfalls in the diagnosis of subarachnoid hemorrhage". *New England Journal of Medicine* 342.1 (2000): 29-36.

5. Leach JL., *et al*. "Imaging of Cerebral Venous Thrombosis: Current Techniques, Spectrum of Findings, and Diagnostic Pitfalls 1". *Radiographics* 26.1 (2006): S19-S41.

6. Van De Bruaene A., *et al*. "Iron deficiency is associated with adverse outcome in Eisenmenger patients". *European Heart Journal* 32.22 (2011): 2790-2799.

7. Vongpatanasin W., *et al.* "The Eisenmenger syndrome in adults". *Annals of Internal Medicine* 128.9 (1998): 745-755.

8. Attar H., *et al.* "Cardioembolic Stroke in Adults With a History of Congenital Heart Disease". *Stroke* 47.5 (2016): e79-81.

9. Assi TB and Baz E. "Current applications of therapeutic phlebotomy". Blood transfusion 1 (2014): s75-83.

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