Dide Botcazo Syndrome: An Atypical and Rare Presentation of Stroke
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Introduction
Infarcts in the territory of the posterior cerebral artery occur in around 5-10% of all ischemic strokes [1, 2]. Dide-Botcazo syndrome is a rare neuropsychological disorder characterized by the association of cortical blindness and anosognosia with amnesia and topographical disorientation [3]. It is secondary to bilateral occlusion of the posterior cerebral arteries [4].

Thrombolysis in the acute phase is as effective after a PCA infarction as after a stroke in the anterior territory [3]. Mortality after PCA stroke is not significant, but long-term cognitive and behavioral deficits are major [1]. This is a largely unrecognized syndrome. Diagnosis of infarction is difficult, especially in the presence of behavioral disorders. Computed tomography (CT) or magnetic resonance imaging (MRI) confirms infarction in areas of the posterior cerebral arteries territory [4, 5].

We report the case of a man admitted for management of a complete Dide-Botcazo syndrome evolving for one week. Cerebral CT showed bilateral ischemia of the posterior cerebral artery (PCA) territory.

Case Report
We report the case of a 65-year-old patient who was admitted for cortical blindness associating antero-retrograde amnesia, topographical anosognosia as well as aggressive behavior evolving for one week. There was no history of hypertension or significant family history. The medical history included diabetes and previous smoking. On admission, baseline NIHSS score was 9, blood pressure and ECG were normal. Clinical examination revealed right hemiparesis. No other neurological signs or abnormal mental symptoms were noted. Initial laboratory work-up showed elevated LDL levels. Transthoracic echocardiography was normal right. Cerebral CT showed ischemic stroke in the territory of the bilateral posterior cerebral arteries (Figure 1).

The patient was treated with antiplatelet drugs, early rehabilitation and risk factor management. Significant improvement in motor deficit and confusion was observed, but cortical blindness and anosognosia remained permanent.

Fig 1: Bilateral complete infarction of the posterior cerebral artery territory in a 65-year-old man

DISCUSSION

The posterior cerebral artery (PCA) plays a major role in cerebral blood perfusion over a vast territory derived from embryological mesencephalic, diencephalic and telencephalic structures. PCA infarcts manifest themselves through a wide range of symptoms and clinical signs. PCA infarcts account for around 5-10% of all ischemic strokes [1-5]. Posterior cerebral artery occlusion can result in a variety of clinical manifestations, including blindness, confusion and amnesia. Embolism is most common; local atherothrombotic stenosis or occlusion of the PCA is rare. Dide and Botcazo first described the combination of cortical blindness, spatial disorientation and amnesia in 1902 [3].

Rare cases of Dide-Botcazo syndrome have been published [6]. The association of cortical blindness and amnesia is recognized as reflecting the joint contribution of the visual cortex, posterior limbic cortex and thalamus via the posterior cerebral artery. Bilateral lesions are required to induce severe or persistent amnesia through damage to the hippocampus or limbic cortex. Bilateral infarcts of the PCA, including its superficial segments, can also explain cortical blindness [7, 8].

Cortical blindness is the consequence of bilateral destruction of the striate cortices or of the optic radiation underlying the cortices. Topographicalagnosia reflects damage to the parahippocampal locus area or to connections between this area and the visual cortex [9]. Anterograde amnesia is probably linked to hippocampal and thalamic lesions, and to damage to limbic white matter pathways caused by bilateral posterior strokes [6-8].

CONCLUSION

Dide-Botcazo syndrome is a very rare condition characterized by the combination of cortical blindness and anosognosia with amnesia and topographical disorientation, secondary to bilateral occlusion of the posterior cerebral arteries. However, it is important to consider cerebral infarction in the presence of the combination of cortical blindness and amnesia, which are atypical symptoms of a stroke, leading to a delay in diagnosis and immediate management.

Competing Interests: None

Patient Consent: Written and informed consent for publication of the case was obtained from the patient.

REFERENCES


5. Brandt, T., Steinke, W., Thie, A., Pessin, M. S., & Caplan, L. R. (2000). Posterior Cerebral Artery Territory Infarcts: Clinical Features, Infarct Topography, Causes and Outcome 1 The authors dedicate this paper to Michael S. Pessin, MD, who died before publication. Multicenter Results and a Review of the Literature. Cerebrovascular Diseases, 10(3), 170-182.


