



Differentiation between Call Fleming syndrome and nervous system vasculitis

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Introduction

Call Fleming syndrome is a reversible cerebral vasoconstriction syndrome of idiopathic etiology that causes thunderclap headache. It is a condition with a good prognosis, but clinically and radiologically similar to other diseases, such as nervous system vasculitis.

Objectives

To bring clinical, radiological and laboratorial aspects of Call Fleming syndrome and nervous system vasculitis and provide the differentiation of both.

Methodology

The research is an integrative review, carried out in June 2022, using the SciElo and Pubmed databases. From the keywords: Call-Fleming syndrome) AND (Diagnosis), (Case report), (Radiology), the clinical, laboratorial and radiological profile of the Call-Fleming Syndrome was traced while the keywords (Nervous System) AND (Vasculitis) AND (Diagnosis), (Case report), (Radiology) were used to CNS vasculitis. Then, from case reports, radiology articles and laboratory analyses, the diseases in question were compared in terms of similarities and differences.

Results

Both pathologies are similar from a clinical point of view: they are frequent causes of thunderclap headache, a sudden and severe headache that reaches its maximum intensity within 1 minute. Some patients with Call-Fleming syndrome may present several neurological deficits, the more frequent in case reports are hemiparesis, hemianopia, and loss of balance. The disease has a limited course that lasts from 1 to 3 months, however, its main complications are hemorrhages and cerebral infarction. The age group affected in the reported cases ranges from 10 to 76 years and there is a slight predominance of cases in women.

As for vasculitis, the onset of the condition can be sudden or insidious, in addition to neurological deficits, cognitive deficits such as confusion, lethargy, dementia and psychiatric disorders are described in most case series. Convulsive manifestations may also be present. As possible complications, vasculitis are related to recurrent transient ischemic stroke and hemorrhages. Chronic headaches and episodes of ischemia in younger patients are typical of vasculitis.

As seen, the picture of both diseases is similar. The simplest differentiation is in case of systemic vasculitis cause there are dermatological and rheumatological signs such as livedos reticularis, arthralgias, petechiae, etc. In Call-Fleming syndrome, CSF is normal or with small changes, such as leukocytes between 5 to 35 uL and red blood cells. In vasculitis, in 90% of patients, CSF analysis is abnormal with elevated lymphomonocytic pleocytosis or protein and oligoclonal bands may be present.

The CT scan is normal in most cases of Call Fleming without associated hemorrhages or infarction. There is presence of multifocal segmental vasoconstriction of cerebral arteries in the angiotomography or MRI, but the findings disappear in the angiography about 12 weeks after the onset of the condition.

It is unlikely that a patient with a normal MRI will suffer from vasculitis. A tumor-like lesion on MRI can be found in primary vasculitides of the nervous system, but the most common findings are segmental narrowing in small and medium-sized arteries and lesions of ischemia. Angiography also shows narrowing and a way to differentiate from Call-Fleming syndrome is the nimodipine test during angiography, which demonstrates the reversibility of constrictions.

Conclusion

The diagnosis of vasculitides of the nervous system and Call Fleming syndrome are challenging and complex due to the similarity with other diseases. Both a false positive and a false negative diagnosis of vasculitis can potentially lead to a fatal outcome, so diagnostic accuracy is critical. The main ways to differentiate a case of Call Fleming syndrome from vasculitis is by analyzing the CSF, which is more likely to demonstrate pleocytosis or increased protein in vasculitis, and also by the nimodipine test during angiography.

Keywords: Call Fleming syndrome, Vasculitis, Nervous system, Thunderclap headache.