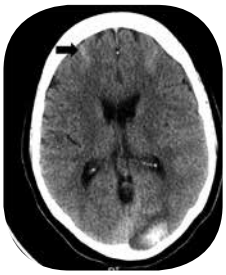

Reversible Cerebral Vasoconstriction: Subarachnoid Convexity Hemorrhage

Síndrome de vasoconstricción cerebral reversible: Presentación como hemorragia subaracnoidea de la convexidad

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Key words (MeSH)
Vasospasm, intracranial
Vasoconstriction
Subarachnoid hemorrhage

Summary

Objective: The purpose of this study is to describe the clinical, epidemiological and imaging characteristics of 5 patients with a diagnosis of reversible vasoconstriction syndrome. **Methods:** A retrospective evaluation of the clinical records of 5 patients with clinical and imaging diagnosis of reversible cerebral vasoconstriction syndrome was carried out in a high complexity hospital in the city of Medellín. **Results:** The five cases were women with an average age of 50 years. In one case, the consumption of energy drinks with a high taurine content was identified as a possible causative agent. All cases presented with brain convexity subarachnoid haemorrhage. The clinical outcome was favorable. **Conclusion:** The reversible cerebral vasoconstriction syndrome should be included in the differential diagnosis of convexity subarachnoid haemorrhage. It occurs predominantly in women between the fourth and sixth decade of life and usually has a benign course.

Palabras clave (DeCS)
Vasoespasmo intracraneal
Vasoconstricción
Hemorragia subaracnoidea

Resumen

Objetivo: El propósito de este estudio es describir las características clínicas, epidemiológicas e imaginológicas de 5 pacientes con diagnóstico de síndrome de vasoconstricción reversible. **Métodos:** Se hizo una evaluación retrospectiva de las historias clínicas de cinco pacientes con clínica e imágenes compatibles con síndrome de vasoconstricción cerebral reversible en un hospital de alta complejidad de la ciudad de Medellín. **Resultados:** Los cinco casos correspondían a mujeres con una edad promedio de 50 años. En un caso se identificó como posible agente causal el consumo de bebidas energizantes con alto contenido de taurina. Todos los pacientes se presentaron con hemorragia subaracnoidea en la convexidad y evolucionaron de forma favorable. **Conclusión:** El síndrome de vasoconstricción cerebral reversible debe ser uno de los diagnósticos diferenciales etiológicos de hemorragia subaracnoidea de la convexidad; se presenta predominantemente en mujeres entre la cuarta y sexta década de la vida y usualmente tiene un curso benigno.

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Introduction

The syndrome of reversible cerebral vasoconstriction is described as a clinical and radiological entity characterized by severe headache and segmental vasoconstriction of cerebral arteries, which may have a benign course or be accompanied by intracranial hemorrhage or ischemic events, resulting in neurological deficit (1). The purpose of this study is to describe the clinical, epidemiological and imaging characteristics of five patients with a diagnosis of reversible vasoconstriction syndrome.

Methods

A retrospective evaluation was made of the histories of five patients with clinical and images compatible with reversible cerebral vasoconstriction syndrome in a high complexity hospital in the city of Medellín.

The diagnosis was made under the following criteria (1):

» Acute headache, with or without associated neurological deficit.
» No new symptoms after one month from the beginning of the clinical picture.
» Without aneurysmal subarachnoid hemorrhage.
» Examination of cerebrospinal fluid (CSF) with proteins <80 mg / dl; leukocytes <10 mm^3 and normal glucose levels.
» Multifocal segmental cerebral vasoconstriction demonstrated by arteriography, angiotomography or magnetic resonance angiography.
» Reversibility of angiographic abnormalities in the 12 weeks after the start of the clinical picture.

All patients underwent magnetic resonance imaging (MRI), simple and with skull contrast medium that included angiographic phase. Laboratory data were analyzed, including rheumatological profile, to rule out primary or secondary vasculitis of the central nervous system, and CSF examination in two cases, to rule out an inflammatory process.

The data were obtained by reviewing the medical records and phone calls to the patients during the clinical follow-up.

Results

The five patients were Hispanic women with an average age of 50 years. None had a history of high blood pressure. Four of them presented to the emergency department with headache "in thunder". One patient presented with seizures and encephalopathy. All episodes of headache "in thunder" began at rest. The maximum pain intensity reported by the patients was 10/10 on the analogous pain scale. One patient, who had a clinical history of migraine, indicated that the pain was of clinical characteristics completely different from the headache she usually suffered (table 1).

Table 1. Symptoms of patients

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age	53	55	33	59	50
Presentation	Headache "in thunder"	Headache "in thunder"	Headache "in thunder"	Headache "in thunder"	Seizures, encephalopathy
Physical examination	Bradipsychic Disoriented Support for ambulation	Without deficit	Without deficit	Without deficit	Bradipsychic
Background	Migraine managed with ergotamine	No	Consumption of 4 to 5 cans of energizers per day	No	No
SBP* > 140 mm Hg on admission	No	No	No	No	No
Autoimmunity	No	No	No	No	No
Ischemia	No	No	No	No	No

*SBP: Systolic Blood Pressure.
Source: Self made.

No patient had systolic blood pressure above 140 mm Hg on admission to the emergency department. One patient was disoriented and bradipsychic, another patient had seizures and encephalopathy, the other three had no neurological deficit.

In one case, the consumption of energy drinks with high taurine content was identified as a possible causative agent. In all cases, the disease manifested with subarachnoid hemorrhage of the convexity, without evidence of aneurysmal bleeding (figure 1).

In all cases, autoimmune causes were ruled out. Antinuclear antibodies, anti-Ro, anti-La, anti-SM and anti-RNP, lupus anticoagulant, anticardiolipin antibodies were requested, which were negative.

In two of them lumbar puncture was performed, but no proteinuria or pleocytosis was found in the cerebrospinal fluid. A patient underwent meningeal and temporal artery biopsy, with which vasculitis was ruled out.

All patients progressed favorably, with clinical improvement and resolution of vasoconstriction at 12 weeks, evidenced by control image (figure 2). In none of the cases were there associated ischemic events.

Discussion

Reversible cerebral vasoconstriction syndrome is a clinical and radiological entity that is characterized by high intensity headache and segmental vasoconstriction of reversible cerebral arteries, which usually resolves in a period of 3 months (1-5).

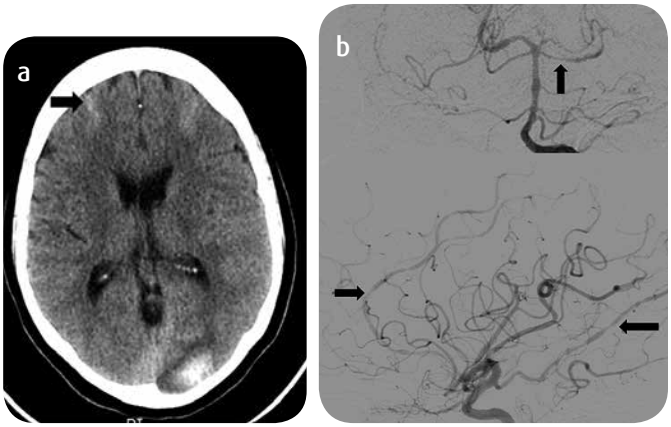

Figure 1. a) CT of simple skull: subarachnoidal hemorrhage in the convexity. b) Arteriography: bleeding of aneurysmal origin is ruled out and a diffuse decrease in vascular diameter is observed in the posterior cerebral arteries, superior cerebellum, posterior communicator, middle cerebral and pericallose arteries.

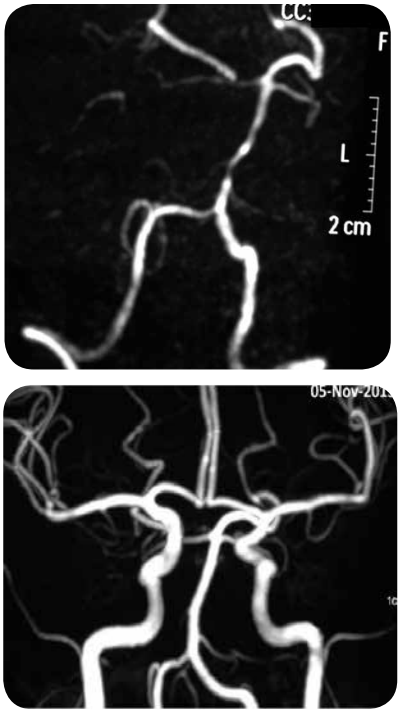

Figure 2. a) MRI: vasoconstriction of posterior circulation. b) Control MRI at 14 weeks: reversibility of initial angiographic abnormalities is observed.

This syndrome is not a single entity, it should be considered the common manifestation of multiple disorders characterized by reversible vasoconstriction of cerebral circulation (6).

The term reversible cerebral vasoconstriction syndrome groups different clinical entities that include Call Fleming syndrome, "thunder" headache, postpartum angiopathy, drug-induced angiopathy, postcoital headache, migraineous vasospasm, and benign angiopathy of the central nervous system (7,8).

Its actual incidence is uncertain, but recent reports indicate that this entity is underdiagnosed (3,4). It mainly affects patients between 30 and 50 years of age, with a mean presentation between 42 and 45 years of age, with a male-female ratio of 1: 2.4 (1).

Reversible cerebral vasoconstriction syndrome can occur spontaneously, but up to 50% of cases are associated with the use of vasoactive drugs or the postpartum state. Sympathomimetic medications usually used to treat the symptoms of respiratory diseases, such as phenylpropanolamine and pseudoephedrine, as well as anti-migraine medication, have been associated with subarachnoid hemorrhage and reversible cerebral vasoconstriction syndrome (1,3,5,9).

The key mechanism of reversible cerebral vasoconstriction is the alteration in vascular tone, which is probably caused by sympathetic hyperreactivity, endothelial dysfunction and oxidative stress, conditions that are required to generate the severe vasoconstriction produced by the clinical symptoms of these patients. The association of reversible cerebral vasoconstriction syndrome with the increase in blood pressure, ingestion of sympathomimetics and pheochromocytoma support the participation of sympathetic hyperreactivity in its pathogenesis. On the other hand, the overlap between reversible posterior encephalopathy syndrome and reversible cerebral vasoconstriction syndrome supports the importance of endothelial dysfunction in the pathogenesis of this entity (1,3,10-12).

Reversible cerebral vasoconstriction syndrome may be associated with seizures, encephalopathy, focal neurological deficits, altered mental status, transient ischemic attacks, ischemic cerebrovascular events, intracranial hemorrhage, and cerebral edema (1,13).

Cortical subarachnoid hemorrhage occurs in 30 to 34% of cases. The rupture of small pial vessels, due to the lack of coordination of the vascular tone, explains the subarachnoid hemorrhage, which usually occurs in the convexity and must be differentiated from the hemorrhage produced by amyloid angiopathy, PRES syndrome, trauma, septic aneurysms, vasculitis, coagulopathy and superficial vascular malformations (4,5,14,15).

The diagnostic criteria are those proposed by the International Headache Society (1), and although they have not been validated in a prospective study, they are quite useful from the clinical point of view:

- Acute, severe headache, with or without associated neurological deficit.
- No new symptoms after one month from the beginning of the clinical picture.
- Without aneurysmal subarachnoid hemorrhage.
- CSF with proteins <80 mg / dl; leukocytes <10 / mm^3 and normal glucose levels.
- Multifocal segmental cerebral vasoconstriction demonstrated by arteriography, angiotomography or magnetic resonance angiography.
- Reversibility of angiographic abnormalities in the 12 weeks following the onset of the clinical picture.

One of the main differential diagnoses is vasculitis of the central nervous system. Patients with primary angiitis of the central nervous system usually have a fulminating course with poor prognosis, in case immunosuppressive therapy is not initiated early. Primary angiitis of the central nervous system is prevalent in elderly men and it is characterized by a headache of a progressive nature; cerebrospinal fluid contains proteins greater than 100 mg / dl and greater than 5-10 cells/ mm, and initial MRI shows multifocal infarcts in most cases (2,6,16).

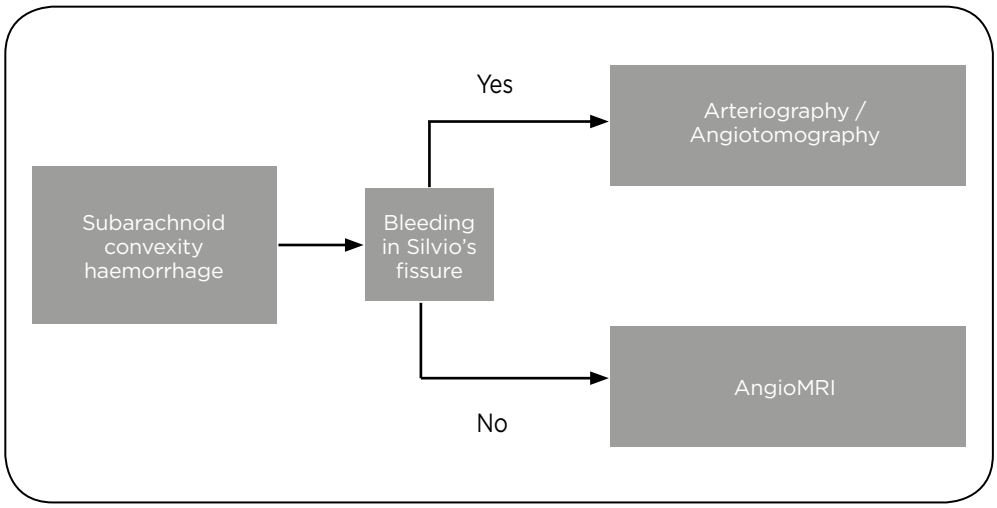

Figure 3. Diagnostic algorithm. In the case of subarachnoid hemorrhage in Silvio's fissure, as a first possibility. Aneurysmal subarachnoid hemorrhage should be ruled out and the study continued with arteriography or angiotomography.

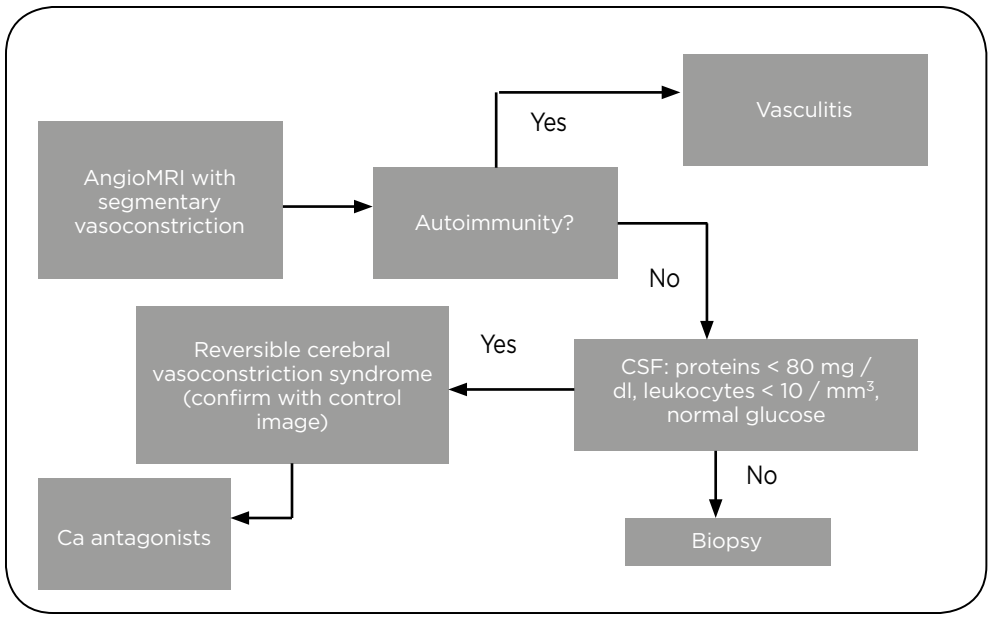

Figure 4. Diagnostic algorithm. Reversible cerebral vasoconstriction is a diagnosis of exclusion and if segmental vasoconstriction is found in angiography, initially central nervous system vasculitis should be ruled out, by means of paraclinics that determine rheumatologic disease, and cerebrospinal fluid that indicates inflammatory disease.

Considering the cases presented in this study and the manifestation as subarachnoid hemorrhage of the convexity, the following diagnostic algorithm is proposed (figures 3 and 4).

Temporal artery biopsy is not recommended for the diagnosis of reversible cerebral vasoconstriction syndrome and should only be performed in cases in which cerebral angiitis is highly probable (7,17).

All patients require symptomatic management. The triggering factor must be identified and eliminated; administer analgesics, medication for the control of blood pressure and anticonvulsant prophylaxis. Activities that generate an increase in symptoms, such as Valsalva maneuver, physical exercise and sexual activity should be avoided. Calcium blockers, including nimodipine, have been administered in different studies, but a change in the evolution of cerebral vasoconstriction has not been demonstrated. Intraarterial administration of vasodilators and balloon angioplasty have been described in severe cases, but its indication is unclear and occasionally vasoconstriction recurs. Some studies suggest that the use of steroids may be deleterious to the patient (1).

The prognosis for most patients is good. The syndrome is usually monophasic with resolution of symptoms in the first three weeks. The resolution of vasoconstriction occurs in the first 12 weeks. 5 to 10% of cases are associated with neurological disability and mortality. Most of these cases of poor prognosis occur in the context of reversible cerebral vasoconstriction syndrome in the postpartum period (1,7).

Conclusion

The syndrome of reversible cerebral vasoconstriction must be one of the differential etiological diagnoses of subarachnoid hemorrhage of the convexity; It manifests predominantly in women between the fourth and sixth decade of life and usually has a benign course.

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