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Central nervous system vasculitis secondary to sarcoidosis presenting with hemorrhagic stroke

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Description

A sixty-two year old woman, under treatment since five years for lung sarcoidosis, presented in the ER of our facility with severe thunderclap headache and hypertensive crisis. The ongoing corticoid therapy (twenty milligrams of oral prednisolone daily), well tolerated before, was discontinued and the patient underwent an unenhanced brain CT scan.

The imaging revealed a right parietal hematoma, and the patient was hospitalized for conservative treatment. Three days later her condition deteriorated substantially with meningism, neck stiffness, photophobia and headache. A follow-up CT and, eventually a brain MRI, documented bilateral parietal hematomas of different size and chronology (Figure 1).

Assuming the etiology related to the background, long-term disease (sarcoidosis), we performed an angiography of blood vessels. The obtained images were highly suggestive for a vasculitis, with multiple narrowing of distal, small-caliber vessels in the territories of cerebral media arteries, bilaterally (Figure 2).

Vasculitis is a blood vessel walls inflammatory disease leading to destruction of normal morphologic and physiological features of vessels, which results in segmental narrowing to occlusion and/or creation of micro-aneurysms. Hemorrhage or ischemia might follow [1]. There are numerous forms of vasculitis affecting the central nervous system. Primary angiitis of the Central Nervous System (PACNS) is a relatively rare form [2]. PACNS is a rare inflammatory disease predominantly affecting small to medium size vessels, confined only to the CNS without evidence of systemic vasculitis, whereas CNS vasculitis is secondary when it occurs in the presence of a systemic inflammatory disease. Sarcoidosis, as a systemic disease presumably of autoimmune origin, affects CNS (neurosarcoidosis) although reports of a secondary vasculitis in this setting are uncommon [3]. Treatment of vasculitis consists on corticosteroids as first line therapy for the disease itself, and treatment of the complications depending on disease manifestations. When corticosteroids' effect is insufficient, immunomodulatory drug are helpful as second line therapy [4].

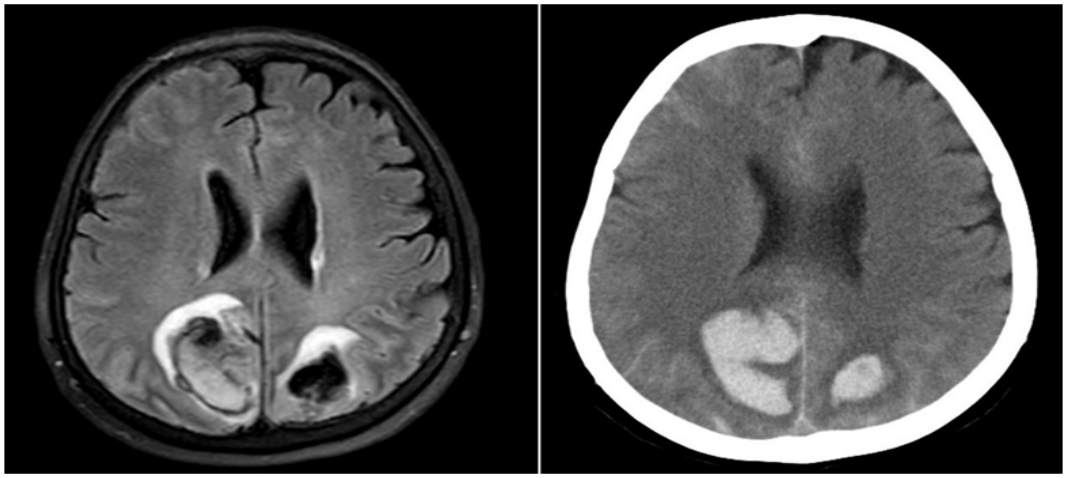

Figure 1: (1A) left inset, T1 flair MRI of brain images showing bilateral parietal hematomas. **(1B)** right inset, unenhanced axial CT images of the same region.

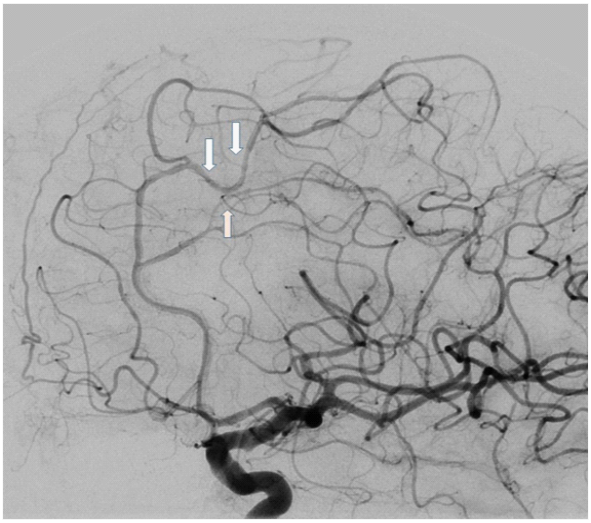

Figure 2: Segmental narrowing of distal, small-caliber vessels in the territory of medial cerebral artery (arrows).

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