

Case Report

Imaging findings of primary angitis of central nervous system: case report and literature review

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Abstract: Primary Angitis of Central Nervous System (PACNS) is the inflammatory disorder of the brain and spinal cord. It is rare, very less studied and usually found in middle aged males. Though, in most cases, radiological findings are nonspecific, they can give very important information to the diagnosis. Literatures suggest that in the case of PACNS, MRI findings are usually bilateral, with involvement of multiple blood vessels, lesions appearing at multiple sites and in different forms. In contrast to these, our case shows involvement of few cerebral blood vessels unilaterally. The vascular lesion is associated with a single mass lesion in the cortex and subcortex, and shows features of edema and hemorrhage. Clinical manifestation of our case is nonspecific and chronically progressive.

Keywords: Primary angitis, central nervous system, imaging

Introduction

Primary angitis of CNS, the inflammatory disorder restricted to brain and spinal cord, is rare and poorly understood. This kind of disorder got a distinct nosological identity in the mid-1950s after the work of Cravioto and Feigin [1]. They described several cases of non-infectious granulomatous angitis associated with the nervous system. Since then, this condition has been given different terms like granulomatous angitis of the CNS, non-infectious or idiopathic granulomatous angitis of the CNS, giant-cell arteritis of the CNS, isolated angitis of the CNS, primary angitis of the CNS, and benign angiopathy of the CNS [1-5].

A very low annual incidence of 2.4 cases per million patients per year has been reported [6]. PACNS accounts for only 1% of the totally vasculitides. Most patients who are suspected to have PACNS actually will present in different disorder [7]. Middle aged people are more likely to be affected than other age groups. In a study conducted in more than 8 patients, an onset median age of approximately 50 years was reported. Incidence among Male is twice as much as in Female [8].

Neurological symptoms

Primary angitis of the central nervous system can be focal and diffuse because of the vasculitis of the intracerebral blood vessel [7]. The symptoms may mimic those of different conditions ranging from angiocentric infections to malignant neoplasms. Long standing headache and altered mental status are the most common symptoms.

The modality of treatment differs vastly in these mentioned conditions. Immunosuppressant therapy which is apt for PACNS could prove to be very harmful to the infective etiologies. Therefore, accurate diagnosis of the condition is crucial in order to execute an appropriate treatment and attain an optimum prognosis [7].

There is no standard radiological test to diagnose PACNS. Brain MRI reveals multifocal abnormal signals in white and gray matter. Leptomeningeal enhancement is less commonly used [9]. MRA is not useful in diagnosing PACNS as it cannot demonstrate inflammatory changes in smaller blood vessels. Cerebral angiography can show ectasia and stenosis. But the sensitivity is only about 60% [5, 10].

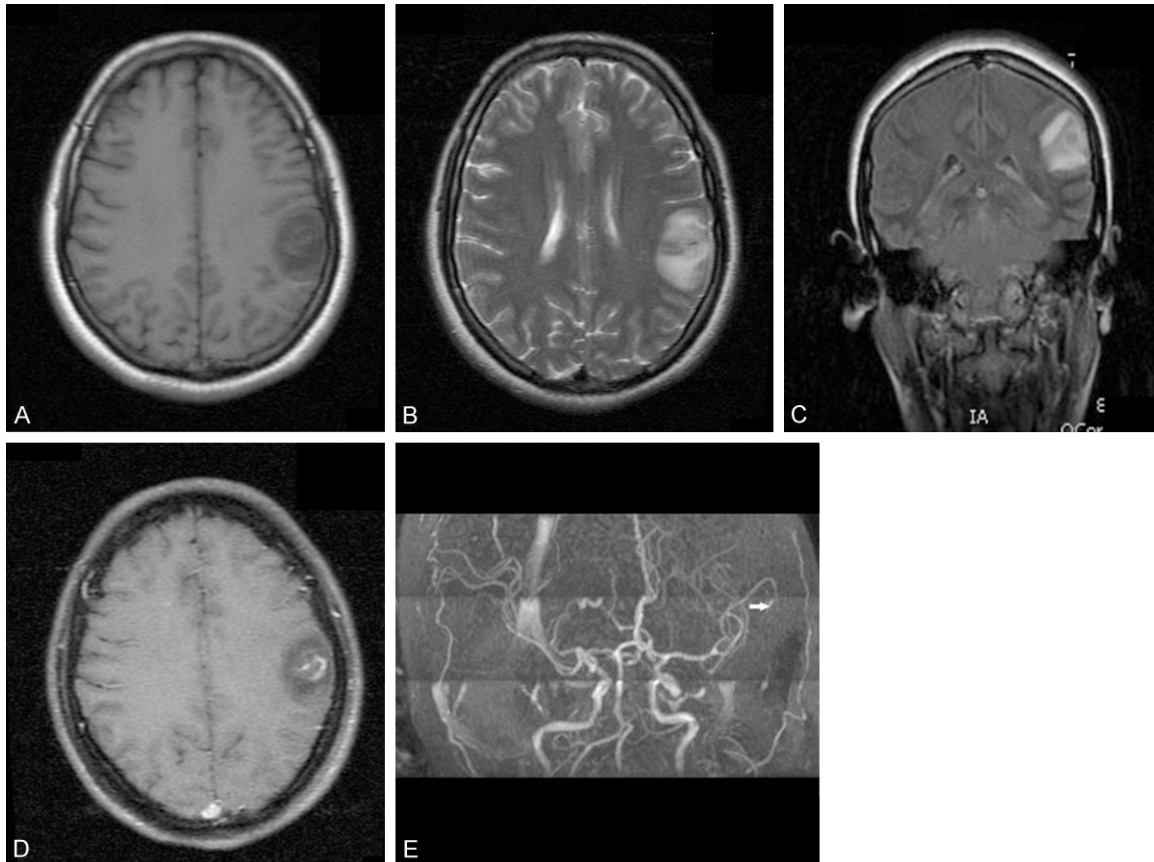


Figure 1. MRI shows a lesion measuring 3.5×2.9 cm. In the fronto parietal region of the left cerebrum with low signal intensity on T1WI, high signal intensity on T2WI and Flair. Contrast enhanced study shows obvious arc like enhancement at the centre of the lesion, suggestive of fresh bleed. The peripheral part remains unenhanced. MRA shows sudden termination of one branch of left middle cerebral artery (as shown by white arrow).

Secondary causes of vasculitides need to be ruled out via blood and CSF examinations. Brain biopsy is still the gold standard for the diagnosis of PACNS. However, even brain biopsy is associated with a negative rate of around 25%. Furthermore, brain biopsy is associated with transient and permanent morbidity of 14% and 4% respectively [11, 12], which increases the significance of imaging modality for probable definitive diagnosis in this condition.

Imaging

For patients with suspected PACNS, MRI should be the neuroimaging modality of choice [7]. MRI findings are abnormal in more than 90% of the patients. But the findings are non specific. The most common abnormalities are detected in the descending order of the subcortical region, the deep gray matter, the deep white matter and the cerebral cortex. Multiple ischaemic foci of different chronology, along with

hemorrhages, leukencephalopathies, and gadolinium-enhanced meninges have all been reported. Such infarcts may be seen in half of the cases. They are usually bilateral in multiple vessel branches. A single tumor-like lesion or isolated myelopathies are further possible MRI presentations [13]. Recently, a cerebral angiography showing findings consistent with inflammation of bilateral large or small vessel is frequently used as the clue for diagnosis of PACNS [14]. But there is a great range of conditions mimicking this kind of vasculitis, such as vasoconstriction associated with migraine, drugs, eclampsia, and hypertension. Mass effects which can give rise to a misleading impression of a malignant neoplasm can be seen in 15% of cases. Some cases show diffuse small vessel changes of ischaemic demyelination. Very rarely, confluent white matter lesion can be seen leading to a mistaken diagnosis of multiple sclerosis [15]. One-third of the cases shows

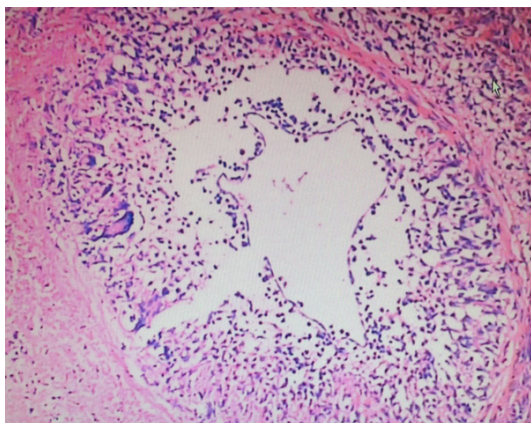


Figure 2. Compromised arterial wall by inflammatory infiltrate of lymphocytes and multinucleated giant cells (HE stain, $\times 100$ magnification).

lesion enhancement on contrast administration. Therefore, tumors, trauma and spasm after sub arachnoid hemorrhage or angiography should also be considered as important differential diagnoses. This case report has been approved by the institutional review board of our hospital with completed patient informed consent.

Case report

A 40-year-old female was presented in the OPD (Out Patient Department) with complain of headache on and off in the last six months. Headache was associated with dizziness. The severity of these symptoms was increasing with time. There was a history of one episode of syncope (half month ago) which occurred at the height of headache and dizziness. She had a past history of nephritis, diagnosed one year ago and was cured after treatment. There was no history of hypertension, heart disease, vascular disease, diabetes mellitus and trauma. There was no history of surgery in the past. Her vital signs were measured to be within normal limits.

MRI findings

MRI showed a lesion measured 3.5×2.9 cm. in the fronto parietal region of the left cerebrum. The lesion showed low signal intensity in T1WI (**Figure 1A**) and high signal intensity in T2WI (**Figure 1B**) and Flair (**Figure 1C**). It was surrounded by an area of hemorrhage and oedema. Owing to oedema, the lesion showed slight mass effect leading to slight distortion of sulci

and gyri patterns in its vicinity. Contrast enhanced study showed obvious arc like enhancement at the centre of the lesion suggesting fresh bleed, and the peripheral part remained unenhanced (**Figure 1D**).

MRA showed sudden termination of one branch of left middle cerebral artery (as shown by white arrow in **Figure 1E**). Thus, in the case we are presenting that the branch of the left middle cerebral artery showed sudden termination most probably due to occlusion by a thrombus. The right part of the brain and right side blood vessels showed no obvious abnormality.

Pathological findings

Pathological report suggested a gross lump in the fronto parietal region with a size of $3 \times 3 \times 2$ cm. Microscopy revealed areas of patchy necrosis with flame like hemorrhages. Blood vessel wall contained numerous inflammatory cells which included lymphocytes and abnormal giant cell reaction. Parts of blood vessels showed thrombosis and necrosis (**Figure 2**). Immunohistochemistry showed CD68 (+), LCA (++), CD38 (+), Ki-67 (=), S100 (+), NF (+), GFAP (+). PAS and Acid fast stain bacteria were negative. The findings were consistent with central nervous system vasculitis.

Discussion

Literatures suggest that the MRI findings in the case of PACNS are usually bilateral [13, 16] with involvement of more than one blood vessel giving rise to appearance of lesions at multiple sites. The lesions vary in location, including subcortical white matter, cortical grey matter and deep grey matter. The lesions appear in different forms, sometimes mimicking a malignant lesion, and sometimes showing a diffuse small vessel changes of ischemic demyelination [7]. However, unilateral lesion with involvement of few vessels has also been reported [17], but such finding is very rare. Our case showed unilateral lesion with involvement of few cerebral blood vessels on the right MCA territory. The vascular lesion was associated with a single mass lesion, located in the cortex and subcortex of left fronto parietal region, showing features of edema and hemorrhage. Rest of the brain showed no apparent lesion. This is a unique feature of our case which, to our knowledge, has not been reported so far.

Immunohistochemistry results were consistent with vasculitis and histopathological examination confirmed the diagnosis. Regarding the clinical features of our case, they were rather non-specific. Similar to the cases reported in previous literatures, the signs and symptoms in our case were also chronically progressive.

Disclosure of conflict of interest

None.

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