World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Check for updates

Primary angiitis central nervous system for young adult with retinitis: Case Report

Yahya Farah *, Almaghribi ISRAE, Mustapha HEMAMA, Nizare EL FATEMI and Moulay Rachid EL MAAQILI

Department of Neurosurgery, Hospital IBN SINA, Rabat, Mohammed V University of Rabat, Morocco.

World Journal of Advanced Research and Reviews, 2024, 24(01), 454–458

Publication history: Received on 15 August 2024; revised on 28 September 2024; accepted on 30 September 2024

Article DOI: https://doi.org/10.30574/wjarr.2024.24.1.2950

Abstract

Primary angiitis central nervous system with multiple cerebral cortex and retinal involvement, it is very rare cases, in the literature, PACNS associated with retinal involvement,

In our case is the case report for difficult diagnostic dilemma, with multiple clinical medical trail, also highlighted the accurate diagnostic of PACNS by STB (stereotaxic Biopsy) histopathological and management.

Keywords: Primary angiitis central nervous system; Retinal; Stereotaxic Biopsy; Histopathological

1. Introduction

Primary angiitis of the central nervous system (PACNS) is a rare and severe disease. It was first described by Harbitz in 1922 as an unknown form of angiitis in the CNS. (1)Isolated vacuities of the central nervous system, primary CNS vasculitis, and isolated angiitis of the CNS are common synonyms for the disease.

PACNS mainly affects younger stroke patients lacking cerebrovascular risk factors. (2) Approximately 3–5% of cerebrovascular events in patients aged <50 years are caused by primary CNS vasculitis(3) The mean age of the initial manifestation of adult PACNS is at the end of the fourth decade(3)In retrospective analyses a similar frequency in male and female patients has been shown(4)

Clinical features at diagnosis are highly variable and nonspecific; classic or pathognomonic clinical symptoms are lacking. Major symptoms of cerebral vasculitis are headache (60%), altered cognition (50%) and focal neurologic deficits (e.g. hemiparesis, ataxia, aphasia, dysarthria and visual disturbances) (2) Furthermore seizures and encephalopathy have been reported. In general, the onset of disease is insidious and slowly progressive, but an acute beginning of symptoms can occur(2)Marked constitutional symptoms such as fever, weight loss and night sweats are less frequent and can be indicative of systemic vasculitis(5)

An extremely low annual incidence of 2.4 cases per million patient/years is reported (4). It was considered a distinct clinical entity in 1959 by Cravioto and Feigin. (6)(The diagnosis remains a challenge, since there are no universally accepted diagnostic criteria and imaging findings may not be specific. Different types of clinical presentation have been described, which makes it even more difficult to identify. Unlike systemic vasculitis, this entity lacks positive autoantibodies.

In our case we report PACNS young adult with retinitis, which is successful diagnosis stereotaxic biopsy and treated corticosteroid and respond well.

* Corresponding author: Yahya Farah

2. Case Report

A 22 year old male patient admitted in the department of the neurosurgery at Avicenna hospital in Rabat for head age associated with ,convulsion partial at left side then generalized tonic colonic seizure ,dysarthria, and visual disturbance for short slightness, for 2 years, no vomiting ,there is no history of trauma ,also he develop hemiparesis after couple of month , and admitted at department of the neurology , he had past history of the vasculopahic retinitis, at 2020 treated by ophthalmologist , after the symptoms worsening and also had history of the treatment for tuberculosis ,and epileptic drugs, at department of the neurology , there is no positive family history , Physically patient weight loss up to 10 kg and no jaundice, cyanosis, pale, he is conscious, Gcs 15/15,well oriented and alert ,hemodynamically and respiratory are stable ,pupil equal and reactive , walking normal, there is facial palsy grade 2 on the left hemi facial, also Motoric dysarthria , the power is 4/5 at the left side upper and lower limbs , sensory are intact , and coordination normal .

All routine blood work up was normal and inflammatory markers ANA/ANCA all negative, tuberculosis test negative.

For radiological assessment; - done MRI cerebral,

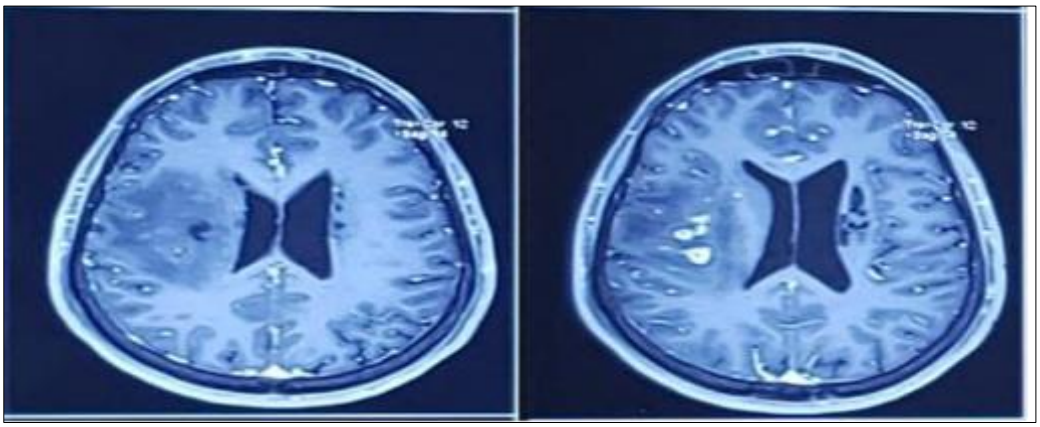
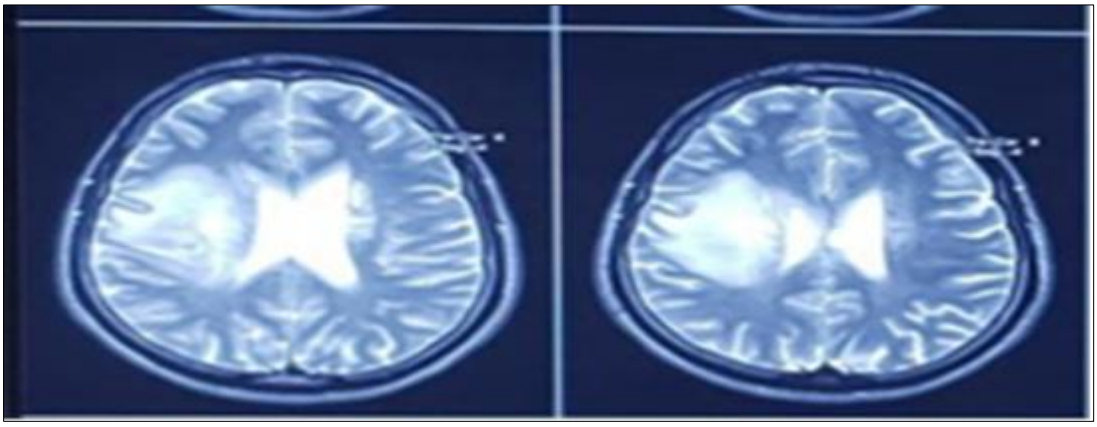
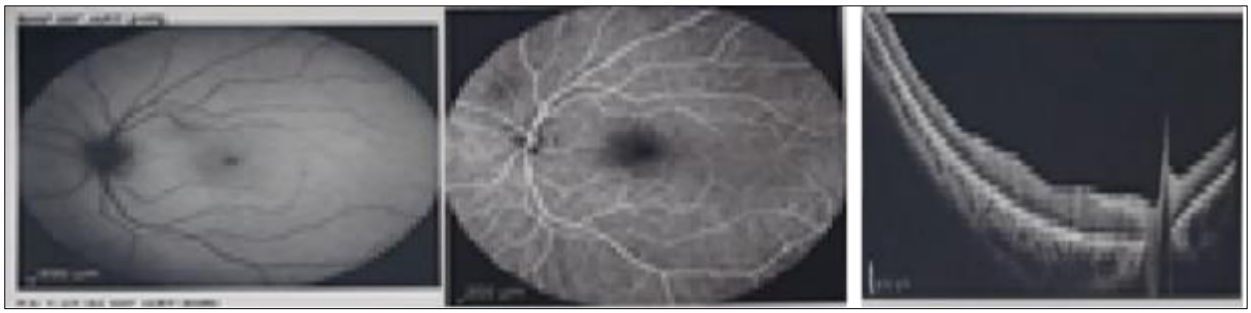


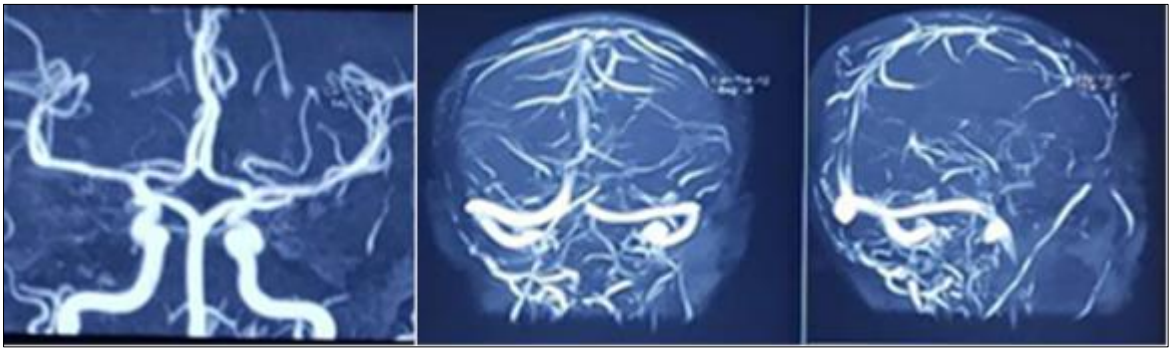
Figure 1 Multiple lesion hyper intense on the cerebral surrounding small edema T1 injected



Finger 2 Flair there massive edema perilesional at right front- parietal

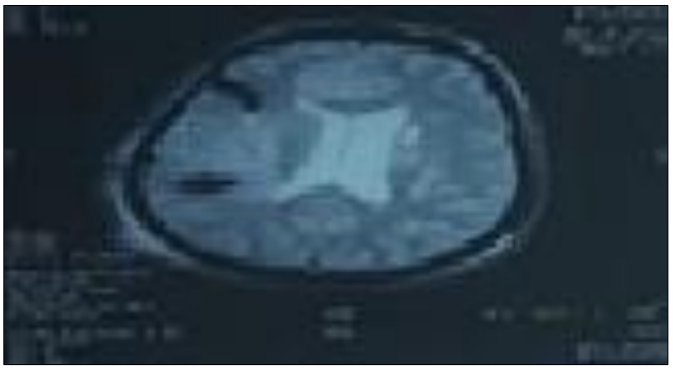


Finger 3 Ophthalmological test, there right side retinal vacuities



Finger 4 Angiography cerebral there is miner lacunar vacuities

Then patient had stereotaxic biopsy (STB) at right side of the cerebral after the histopathological examination confirm lymphocytic inflammatory vasculitis, with gliosis reaction (PACNS) primary angiitis central nerve system .then start treatment for the corticosteroid, immediately after diagnosis.



Finger 5 Post STB control no complication

3. Discussion

PACNS is an entity poorly understood and significant challenges remain for diagnosis and treatment [6]. However, there has been an increased recognition with the diagnostic criteria proposed by Calabrese and Mallek and the number of reported cases have risen substantially. The general advances in diagnosis of these neurological disorders have led to an aggressive diagnostic approach and enriched clinical and pathological descriptions [5,7].

Histologically, the inflammatory process commonly is lymphocytic and affects the medium-sized and small arteries and arterioles of the meninges and cortex of the brain and only rarely the veins and venules. The classic findings of segmental granulomatous vasculitis with multinucleated giant cells occur in less than 50% of patients. Necrotizing vasculitis occurs in 25% of patients. The presence of intimal fibrosis usually signifies healed lesions [8].

The clinical, imaging, and angiography spectra can be very broad. The median age of onset is 50 years, but may affect patients of all ages. The neurological manifestations are diverse, ranging from hyperacute to chronic and insidious. Generally, PACNS consist of headache, altered cognition, focal weakness, or stroke occurring in 30–50% of patients, usually affecting many different vessels [7].

Different patterns and outcomes of the disease have been described, like fulminant-disease onset, spinal involvement [9], prominent leptomeningeal enhancement, and negative cerebral angiography, suggesting that medium-sized vessels are affected. CSF examination reveals evidence for aseptic meningitis in over 90% of patients [10]. Another presentation was described by Molloy et al. [8] characterized by a mass-lesion, making the pathological confirmation essential for diagnosis. Sometimes aggressive immune suppressive treatment could obviate the need for excision [11] At last; there is a group of vasculitis that is characterized by abnormal cerebral angiography and CSF examination but usually a normal brain in the biopsy.

The most important differential diagnosis is the systemic vasculitis, which is characterized by the presence of constitutional symptoms and serological markers indicating systemic inflammation [12]. The hallmark of the PACNS is that inflammatory process is limited to the CNS. Diseases that can mimic PACNS are the reversible cerebral vasoconstriction syndrome, premature intracranial atherosclerosis, fibromuscular dysplasia, moyamoya disease, brain neoplasms, genetic conditions (e.g., CADASIL), posterior reversible encephalopathy syndrome (PRES), chronic hypertension (microvascular cerebral ischemia), demyelinating diseases, and others with multifocal cerebral thromboembolism. It is important to notice that infections (i.e., *Varicella zoster* virus, HIV, and hepatitis C virus) have been associated with cerebral angiographic abnormalities and should be excluded with serological tests [13, 14]. Tuberculosis is another important mimic of PACNS.

No single laboratory test has sufficient sensitivity or specificity to establish the diagnosis, and high clinical suspicion is necessary. Brain biopsy is required to confirm the diagnosis and exclude other causes. Based on published studies, the estimated sensitivity of cerebral angiography for detection of vasculitis is between 27% and 90% and for brain biopsy between 36% and 83% [5, 7]. MRI is the main neuroradiographical modality for the workup of patients with suspected PACNS, and generally, the combination of normal findings on MRI and normal CSF analysis has a high negative predictive value for the diagnosis of PACNS [11].

A common pitfall in the workup diagnosis is to start immunosuppressive treatment without establishment of diagnosis or exclusion of other diseases. Unfortunately, no randomized studies of PACNS have been done, and thus all information on treatment is based on retrospective observational data and clinical experience. The efficacy of most immunosuppressive agents such as azathioprine, methotrexate, and rituximab that are used in systemic vasculitis is still unknown or remains elusive in PACNS [12]. The vast majority of case series suggest a high degree of good outcomes when patients are treated with glucocorticoids or glucocorticoids and cyclophosphamide. However, there are some reports using a therapy combining mycophenolate mofetil and steroids allowing control of the disease with disappearance of the neurological abnormalities, restoration of normal daily activities and dramatic improvement in brain MRI abnormalities [14].

The majority of PANCS reports are from Europe and North America [15]. In Latin America there are no reports of PACNS, and we do not really know the incidence and prevalence of this entity in Colombia. Takayasu arteritis was the most frequently reported form of primary vasculitis overall, from Colombia [16], Brazil, and Mexico ,Even though we think there is some experience with PACNS in Latin America, it has not been published.

In our case report presenting the Importantly to know the clinical heterogeneous presentation of this condition when we handled this group of patients, even the possibility that it has of simulate psychiatric or malignant disorders like from described patients. The high clinical suspicion and appropriate diagnostic approach focused on problems, taking into account unusual entities like this, and it will make one accurate diagnostic and therapeutic approach. We hope that with this case report the medical community is encouraged to report more cases and thus in this way achieve a unanimous concept in respect to the management, diagnostic and therapy of this entity which has a high mortality rate.

4. Conclusion

Primary angiitis central nervous system, which is involved cerebral cortex and retinal most be considers very important differential diagnosis for multiple focal lesion of cerebral, which is mimic to metastatic tumors, infection (tuberculosis), and other inflammatory central nervous system, although extremely rare cases, its clinical important to diagnosis and assessment different clinical presentation, and immediately intervention will improve the outcome of the patient.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

[1] Harbitz F. Unknown forms of arteritis, with special reference to their relation to syphilitic arteritis and periarteritis nodosa. Am J Med Sci 1922; 163: 250–271. [Google Scholar].

[2] Berlit P, Kraemer M. Cerebral vasculitis in adults: what are the steps in order to establish the diagnosis? Red flags and pitfalls. Clin Exp Immunol 2014; 175: 419–424. [PMC free article] [PubMed] [Google Scholar]

[3] Ferro JM. Vasculitis of the central nervous system. J Neurol 1998; 245: 766–76. [PubMed] [Google Scholar

[4] Salvarani C, Brown RD, Jr, Christianson T, et al. An update of the Mayo Clinic cohort of patients with adult primary central nervous system vasculitis: description of 163 patients. Medicine 2015; 94. [PMC free article] [PubMed] [Google Scholar]

[5] Birnbaum J, Hellmann DB. Primary angiitis of the central nervous system. Arch Neurol 2009; 66: 704–709. [PubMed] [Google Scholar]

[6] Cravioto H. and Feigin I., Noninfectious granulomatous angiitis with a predilection for the nervous system, Neurology. (1959) 9, 599–609

[7] Hajj-Ali R. A., Singhal A. B., Benseler S., Molloy E., and Calabrese L. H., Primary angiitis of the CNS, The Lancet Neurology. (2011) 10, no. 6, 561–572, 2-s2.0-79956024180, https://doi.org/10.1016/S1474-4422(11)70081-3.Google Scholar

[8] Molloy E. S., Singhal A. B., and Calabrese L. H., Tumour-like mass lesion: an under-recognised presentation of primary angiitis of the central nervous system, Annals of the Rheumatic Diseases. (2008) 67, no. 12, 1732–1735, 2-s2.0-56749131432, https://doi.org/10.1136/ard.2008.096800.Google Scholar

[9] Goertz C., Wegner C., Brück W., and Berlit P., Primary angiitis of the CNS with pure spinal cord involvement: a case report, Journal of Neurology. (2010) 257, no. 10, 1762–1764, 2-s2.0-79952201362.Google Scholar

[10] Baumer D., Flossmann E., Cudlip S., Quaghebeur G., Jeans A., and Talbot K., Primary angiitis of the CNS mimicking a spinal cord tumour, Journal of Neurology. (2008) 255, no. (11), 1970–1972, 2-s2.0-67149128366, https://doi.org/10.1007/s00415-009-0853-y.Google Scholar

[11] Stone J. H., Pomper M. G., Roubenoff R., Miller T. J., and Hellmann D. B., Sensitivities of noninvasive tests for central nervous system vasculitis: a comparison of lumbar puncture, computed tomography, and magnetic resonance imaging, Journal of Rheumatology. (1994) 21, no. 7, 1277–1282, 2-s2.0-0028040487.Google Scholar

[12] Amara A. W., Bashir K., Palmer C. A., and Walker H. C., Challenges in diagnosis of isolated central nervous system vasculitis, Brain and Behavior. (2011) 1, 57–61.

[13] Cañas C. A., Jiménez C., Arango G. et al., Compromiso vascular en la encefalitis herpética, Revista Facultad de Medicina de la Universidad Nacional de Colombia. (1997) 45, 16–20.Google Scholar

[14] Cañas C. A., Cedeño S. V., Carrascal E., and Vélez J. D., Diátesis trombótica en paciente con infección aguda por citomegalovirus, Acta Médica Colombiana. (2009) 341, 36–139. Google Scholar

[15] Chenevier F., Renoux C., Marignier R., Durand-Dubief F., Hermier M., Streichenberger N., Vukusic S., and Confavreux C., Primary angiitis of the central nervous system: response to mycophenolate mofetil, Journal of Neurology, Neurosurgery and Psychiatry. (2009) 80, no. 10, 1159–1161, 2-s2.0-70349690305, https://doi.org/10.1136/jnnp.2008.154567. Google Scholar

[16] Ochoa C. D., Ramirez F., Quintana G., Toro C., Cañas C., Osio L. F., Cantillo J., Rondón F. et al., Epidemiología de las vasculitis primarias en Colombia y su relación con lo informado para Latinoamérica, Revista Colombiana de Reumatología. (2009) 16, 248–263.