International Journal of Contemporary Pediatrics
Chencheri NC et al. Int J Contemp Pediatr. 2017 Sep;4(5):1886-1889
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20173805

Case Report

A diagnostic dilemma of two rare stroke mimics in a pediatric patient

Nidheesh Cheeyancheri Chencheri[1]*, Pawan Subhash Kashyape[1], Sonia Khamis[2], Maha Mohammed Elamin Agna[1]

[1]Department of Pediatric Neurology, Al Jalila Children's Specialty Hospital Dubai, United Arab Emirates
[2]Department of Pediatric Neurology, Latifa Hospital, Dubai, United Arab Emirates

Received: 29 June 2017
Revised: 18 July 2017
Accepted: 24 July 2017

***Correspondence:**
Dr. Nidheesh Cheeyancheri Chencheri,
E-mail: drnidheesh@gmail.com

ABSTRACT

A seven and half year-old male patient presented with clinical symptoms suggestive of stroke. However, the MRI showed atypical lesions in non-vascular distribution and with varying ages. This led to extensive investigations, which suggested a differential diagnosis of CNS neurosarcoidosis and primary CNS vasculitis. Both these are exceedingly rare conditions needing tissue diagnosis and prolonged immunosuppression.

Keywords: Neurosarcoidosis, Stroke mimics, Vasculitis

INTRODUCTION

A child who presents to the pediatrician's office or emergency department with an acute neurologic deficit poses a diagnostic challenge. Although stroke is one cause of acute neurologic deficits, it is a relatively rare condition, with a reported incidence of 2.1 to 13.1 per 100000 children per year.[1]

Acute neurologic symptoms are frequently attributed to more common alternative diagnoses such as seizure and migraine. In fact, a pediatric stroke team examined their acute stroke calls and found that almost 21% of children had stroke mimics. Among these children, one-third had benign conditions, while two-thirds had serious diagnoses unrelated to stroke.[2] Therefore, it is important that the treating physicians are aware of the wide differentials of conditions that can present like stroke.

We describe a pediatric patient who presented with focal neurological deficit and turned out to have atypical findings in MRI and lab investigations, leading to the differential diagnosis of two extremely rare conditions presenting as stroke mimics.

CASE REPORT

A seven and half years old boy was referred to the neurology department of a tertiary care hospital with history of acute onset of weakness involving right upper and lower limb since, five days. Two weeks back he had history of brief episode of transient loss of vision on the right eye, which recovered within few minutes and for which no medical help was sought. In addition, there was history of transient unsteadiness while walking one and half months back, which resolved spontaneously. This was assumed at the time by his family to be related to school avoidance. There was no history of fever, headaches, loss of consciousness, seizures, trauma or any recent foreign travel.

At the time of admission, he was well looking with normal general examination. There was no pallor,

lymphadenopathy, organomegaly or stigmata of connective tissue disease. Neurologically he had preserved higher functions, speech and cranial nerves. There was grade 3 powers of right lower limb and grade 4 power of right upper limb with spastic catch and brisk deep tendon reflexes on the same side. Left side was normal. There were no abnormal movements or ataxia. Sensory examination was within normal limits. Cardiovascular and respiratory system examination was normal. Thus, the provisional diagnosis was of right sided stroke with preceding history of probable transient ischemic attacks.

Investigations

The patient came with a CT scan done in the referring hospital, which showed an area of hypodensity in the left medial temporal region with associated volume loss suggestive of an old gliotic area. There was also another lesion in the right frontal region which had features of a well circumscribed hyperintense circular lesion with surrounding perifocal hypointensity which appeared like possible calcification or microbleed. These lesions did not correlate with the current clinical findings. He was investigated in our hospital according to stroke protocol.

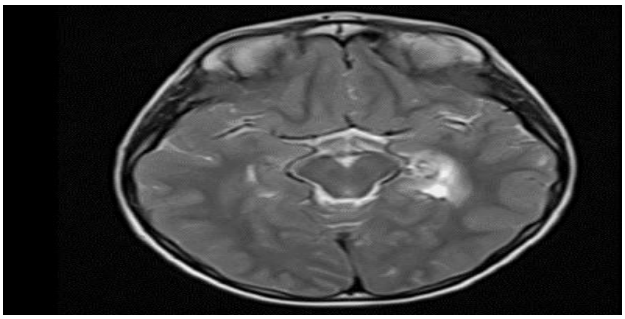

Figure 1: MRI brain-T2 w axial image showing volume loss with gliosis on left temporal regions.

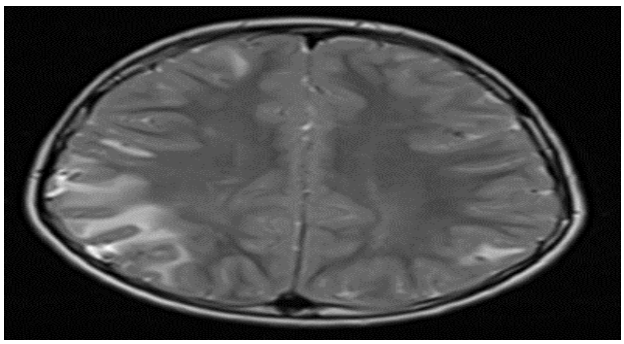

Figure 2: MRI brain T2W axial image showing hyper intensities along subcortical white matter suggesting edema.

Baseline blood tests were normal. MRI brain confirmed the gliotic lesion in the left temporal region, but also showed other lesions in non-vascular distribution consisting of white matter hyperintensities in bilateral subcortical white matter, some with suspicious central hypointense lesions. On gradient echo sequence, there were multiple hypointense lesions with blooming in cortical, subcortical and deep grey matter. Figure 1 to Figure 4 MRI. Thus, the impression was that of multiple areas of microbleeds with surrounding white matter edema (seen as T2W white matter hyperintensities as previously described). Time of flight MRA was normal.

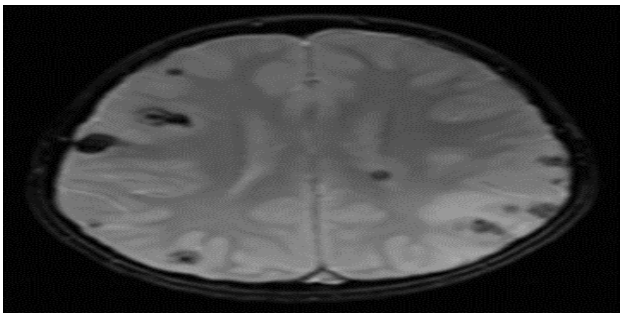

Figure 3: Gradient echo image showing multiple hypointense lesions with blooming in cortical, subcortical and deep grey matter.

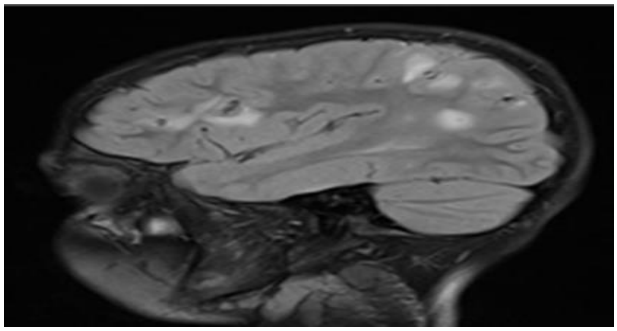

Figure 4: MRI brain T2W saggital image-shows multiple discrete hyperintensities in subcortical white matter.

Autoimmune and vasculitis work up including Anti DS DNA, pANCA, cANCA, phosphatydal serine IGG/IGM, ENA profile, Anticardiolipin IGG/IGM, Compliment C3, C4 was negative. Neurocysticercosis IgG antibody, IgM TORCH titer and T spot for tuberculosis was negative. CSF study showed elevated protein 535mg/dl (10-50), WBC-5 cells and sugar 45 mg/dl (32-82). CSF oligoclonal bands were positive. CSF fungal stain and culture were negative. CSF viral studies and TB PCR were negative.

Considering the MRI findings of multifocal lesions, elevated CSF proteins with positive oligoclonnal bands, possibility of an autoimmune process like neurosarcoidosis or CNS vasculitis was considered. CSF angiotensin converting enzyme (ACE) was sent and was significantly elevated to 14.7 U/L (normal <2U/L).

The patient was further evaluated for systemic evidence of sarcoidosis. CT chest, cardiac Echo and USG abdomen

was normal. Serum angiotensin converting enzyme was normal. The ophthalmology examination showed evidence of significant retinal vasculitis Figure 5.

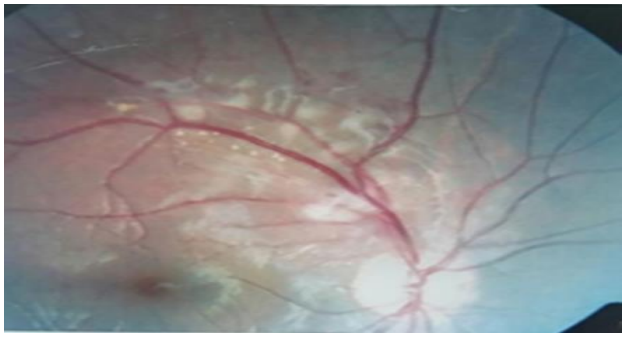

Figure 5: Fundus examination shows exudative vasculitic changes.

Unfortunately, the family did not give consent for tissue diagnosis, which would have required a brain biopsy.

Treatment

The child was started on steroids Methylprednisolone 30mg/kg /day for 5 days and followed by oral prednisolone, with significant clinical improvement. Unfortunately, the family tapered steroids without medical consultation after few weeks. The patient subsequently had a repeat attack of dense hemiplegia affecting the opposite (Left) side, a few days after withdrawal of steroids.

DISCUSSION

When a patient presents with focal neurological deficit, stroke is the top differential diagnosis. However, stroke has several mimics which need to be carefully considered and investigated.[2] The first investigation of choice is MRI of the brain which usually shows involvement of the cortex in a typical vascular distribution in case of arterial ischemic stroke. This is the commonest cause of vascular stroke in young patients.[3] This case proved a diagnostic challenge since the clinical presentation of acute stroke did not match the imaging findings of multifocal lesions of varying ages in non-vascular distribution with perifocal white matter edema, raising instead the suspicion of an ongoing CNS inflammatory/infectious process.

The suspicion was confirmed by the high CSF protein and normal MR angiography which confirmed an inflammatory process and ruled out a large vessel vasculitis or occlusive vasculopathy respectively. Infectious etiologies were ruled out by doing the appropriate investigations, including those for tuberculosis and neurocysticercosis. Also, the patient remained systemically well despite the florid changes on imaging, which made infectious etiology less likely clinically. The lesions on MRI gradient echo sequence raised the possibility of microbleeds or calcifying granulomas. This led to the suspicion of neurosarcoidosis as one of the differential diagnosis. However, the retinal vasculitis and the lack of extra CNS sarcoid involvement also raised the differential diagnosis of primary small vessel CNS vasculitis as another possibility. Thus, based on the clinical course, neuroimaging and lab tests, the two top differential diagnoses were primary neurosarcoidosis causing vasculitis and primary small vessel CNS vasculitis.

Elevated Angiotensin converting enzyme (ACE) in spinal fluid raises the suspicion of neurosarcoidosis due to its high specificity (94%-95%) as in our patient Tissue diagnosis would have helped us to zero in on one of the two, but unfortunately this was not possible.[4] Both primary neurosarcoidosis and primary small vessel CNS vasculitis are exceedingly rare childhood neurological disorders requiring prolonged immunosuppression therapy and with a variable clinical course. They need high index of suspicion for diagnosis. For confirmatory diagnosis, both disorders require tissue diagnosis.[5,6] But this may not be always being possible or consented to by the patient's family due to the invasiveness of the procedure.

However, there are ancillary tests that can guide the physician towards the likely diagnosis. Moreover, the treatment for both conditions is similar.

CONCLUSION

Focal neurological deficits in pediatric patients can have awide differential diagnosis, strokebeing one of them. Not all clinical strokes in children are thromboembolic in nature. In presence of mismatching or unusual imaging findings, one has to think out of the box and consider even extremely rare conditions like neurosarcoidosis and CNS vasculitis.The treatment for these autoimmune disorders is in the form of prolonged immune suppression therapy. The course is highly variable.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Lynch JK, Hirtz DG, DeVeber G, Nelson KB. Report of the National Institute of Neurological Disorders and Stroke workshop on perinatal and childhood stroke. Pediatrics. 2002;109:116-23.
2. Shellhaas RA, Smith SE, O'Tool E, Licht DJ, Ichord RN. Mimics of childhood stroke: characteristics of a prospective cohort. Pediatrics. 2006;118(2):704-9.
3. Carvalho KS, Garg BP. Arterial strokes in children. Neurologic Clinics. 2002;20(4):1079-1100.
4. Khoury J, Wellik KE, Demaerschalk BM, Wingerchuk DM, Cerebrospinal fluid angiotensin-

convertingenzyme fordiagnosis of central nervous system sarcoidosis. Neurologist. 2009;15:108-11.
5. Alrawi A, Trobe JD, Blaivas M, Musch DC. Brain biopsy in primary angiitis of the centralnervous system. Neurology. 1999;53:85-60.
6. Zajicek JP, Scolding NJ, Foster O, Rovaris M, Evanson J, Moseley IF, et al. Central nervous system sarcoidosis-diagnosis and management. QJM. 1999;92:103-17.

Cite this article as: Chencheri NC, Kashyape PS, Khamis S, Agna MME. A diagnostic dilemma of two rare stroke mimics in a pediatric patient. Int J Contemp Pediatr 2017;4:1886-9.