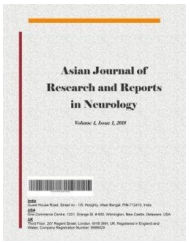

Asian Journal of Research and Reports in Neurology

2(1): 15-20, 2019; Article no.AJORRIN.48263

Unilateral Weakness and Areflexia in a Child Diagnosed with Pseudo-Tumoral Acute Hemi-Cerebellitis

Hanin Al Gethami[1*], Fuad Al Malki[2], Waleed Al-Tuwaijri[2], Duaa Ba-Armah[2] and Ahmed Al Zahrani[3]

[1]Neuro-science Center, King Fahd Medical City, Riyadh, Saudi Arabia.
[2]Department of Pediatric, Pediatric Neurology Division, King Abdullah Children Specialist Hospital, King Abdulaziz Medical City, Riyadh, Saudi Arabia.
[3]Department of Radiology, King Abdullah Children Specialist Hospital, King Abdulaziz Medical City, Riyadh, Saudi Arabia.

Authors' contributions

This work was carried out in collaboration among all authors. Author HAG designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors FAM and WAT managed the analyses of the study. Authors DBA and AAZ managed the literature searches. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Maria Emilia Cosenza Andraus, Adjunct Professor, Universidade Federal do Rio de Janeiro (UFRJ), Brasil.
Reviewers:
(1) Mai Sabry Saleh, National Research Centre, Egypt.
(2) Osvaldo Garrido-Acosta, National Autonomous University of Mexico, Mexico.
(3) Dr. R. Harikumar, Bannari Amman Institute of Technology, India.
Complete Peer review History: http://www.sdiarticle3.com/review-history/48263

Case Report

Received 24 January 2019
Accepted 03 April 2019
Published 17 April 2019

ABSTRACT

Pseudotumoral acute hemi-cerebellitis is an inflammatory condition, a self-limited disease with good prognosis, which does not need treatment in most of the cases. It is very rare worldwide, and few cases have described it before. It has a unique presentation and a characteristic neuroimage finding. The cause is not known however, it can be the primary infection, post-infectious or post vaccination disorder. The present work described that, a child was diagnosed to have pseudo-tumoral acute hemi-cerebellitis with a typical presentation in the form of unilateral weakness and areflexia, which was never been reported before this study.

**Corresponding author: E-mail: hanin_88_jg@hotmail.com;*

Keywords: Areflexia; hemi-cerebellitis; inflammation; post vaccination; immunisation.

1. INTRODUCTION

Pseudotumoral cute hemi-cerebellitis is an inflammatory condition, and it is extremely rare in children, in fact, few cases reported it before. It is clinically present with headache, meningism and sometimes cerebellar signs. MRI appears with focal or diffuse signal changes in one cerebellar hemisphere with or without cerebellar swelling that may look like a tumour radiologically [1,2]. The aetiology is unknown, it can be caused by a primary infection, post-infectious or post vaccination disorder. The most common infectious agents are varicella, measles, mumps and rubella [3,4]. It has a benign course and a significant clinical improvement that has been noticed even in the absence of treatment, with an excellent regression radiologically [2]. However, a few cases were reported to undergo a surgical intervention due to brainstem compression or acute hydrocephalus [5-7].

It is an important differential diagnosis that needs to be considered in the clinical practice because the management is different. Acute cerebellar ataxia and acute cerebellitis are another entity with an overlap between both of them. The acute cerebellar ataxia is characterised by truncal, appendicular, and gait ataxia, nystagmus, dysarthria, and hypotonia. It happens suddenly and resolves over time with normal neuro-image. Unlike acute cerebellitis, it is more severe with less favourable prognosis and abnormal neuro-image, clinically it appears with altered level of consciousness, hydrocephalus, raised intracranial pressure, and focal neurological deficit [8]. In fact, the pseudo- tumoral acute hemi-cerebellitis has been considered as a variant of acute cerebellitis but unilateral.

2. CASE PRESENTATION

A four years old girl medically free presented to ER with irritability, vomiting, fever and right-side weakness for seven days. The vomiting and the fever continued for three days then resolved and the right-side weakness started affecting her upper and lower limbs, she could not bear weight by herself, only with support. The weakness over four days was progressing, then became static. Reaching the point that she cannot raise her arm above her shoulder. She was most of the time crying, refuse anyone to come close to her, not distracted by her toys as before. The following aspects of history were unremarkable including upper respiratory tract infection symptoms, contact with the sick patient, or animals, abnormal movement, or loss of consciousness, cyanosis or shortness of breath, faecal or urinary incontinence, trauma. Her immunisation: was complete. Developmentally, she had normal motor and cognitive function for her age. Nutritional, allergy and family history were unremarkable.

On examination: Vital signs were all normal. She looked well, not in distress but irritable. There were no meningeal signs. Her cranial nerve examination was normal. Her motor examination showed: Upper limb right: 3/5, Upper limb left: 5/5, Lower limb right: 4/5, Lower limb left: 5/5, her tone was normal in the left side, but reduced in the right side. There was an absent knee reflex bilateral and an equivocal plantar response, her gait was unsteady with drift to the right side, the sensory examination was unremarkable.

The differential diagnosis included meningitis, vasculitis, arterial stroke, deep venous thrombosis, tumour, and ADEM (Acute disseminated encephalomyelitis).

The patient was admitted and a full septic workup was done including lumbar puncture. Then she started on antibiotics. Over the whole period of admission, she was vitally stable and afebrile. Later, her cultures came to be negative, so the antibiotics were discontinued.

NCS was normal, MRI brain and spine showed unilateral right cerebellar abnormal FLAIR and T2 signal intensity with cortical swelling. No enhancement post-contrast administration. Most likely represented pseudo-tumoral hemi-cerebellitis (Fig. 1).

The patient had significant clinical improvement with help from physiotherapy and occupational therapy.

Her other parts of investigations like;

CBC, chemistry, renal profile, liver profile, ESR, CRP, ASOT, CPK, toxicology screen, all were unremarkable, CSF showed normal white blood cell, protein, and glucose. The viral study from blood was negative for Measles, Rubella, hepatitis, Varicella-zoster virus, Brucella abortus/ Brucella melitensis, Cytomegalovirus, Epstein–Barr virus, Herpes simplex virus one and two. Viral study from nasopharyngeal aspirate was also negative for Influenza A Virus, Influenza B

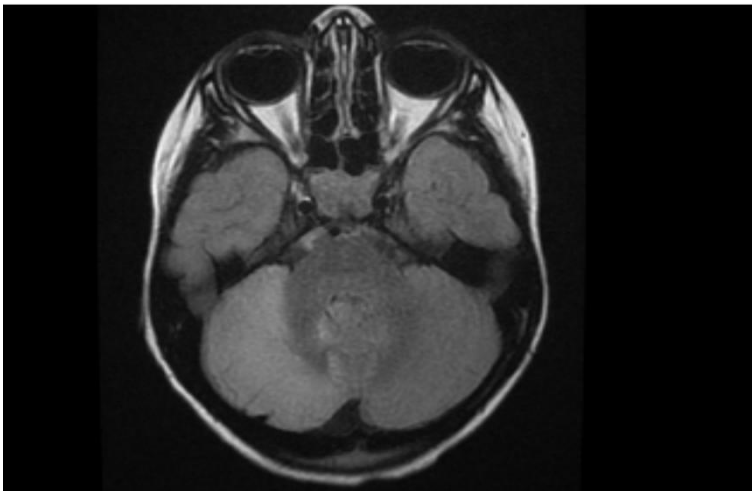

Fig. 1A. Axial T2 Flair

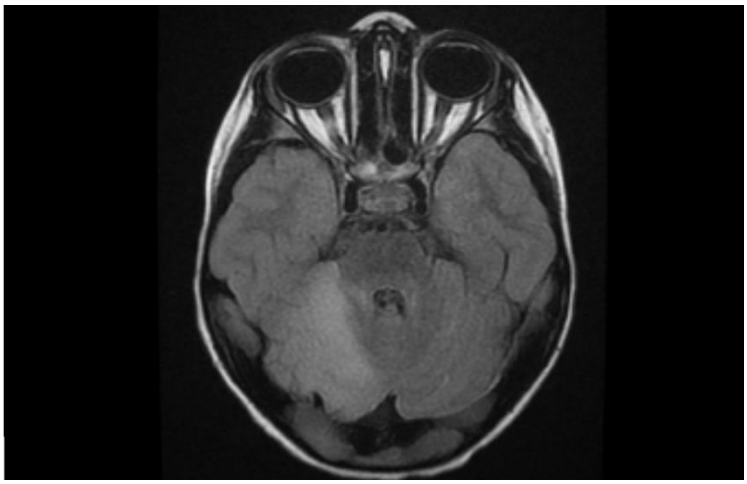

Fig. 1B. Axial T2 Flair

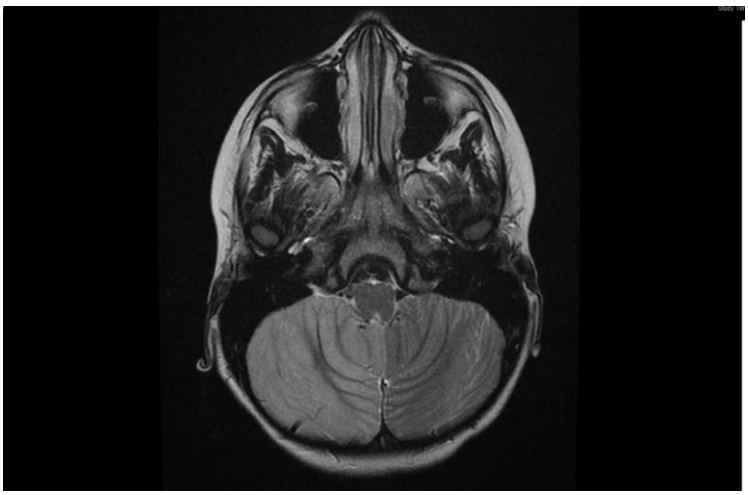

Fig. 1C. Axial T2

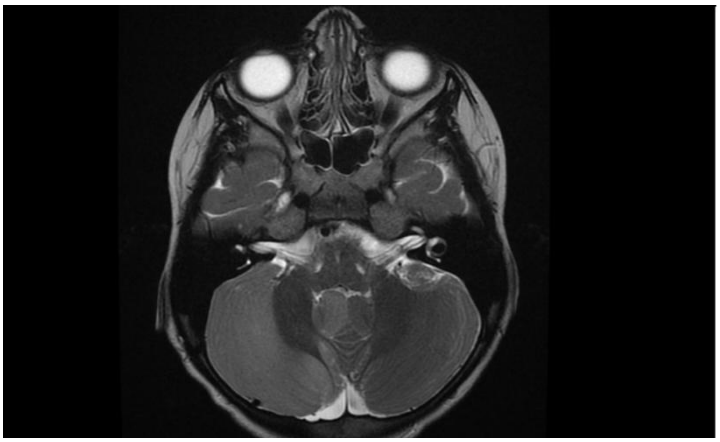

Fig. 1D. Axial T2

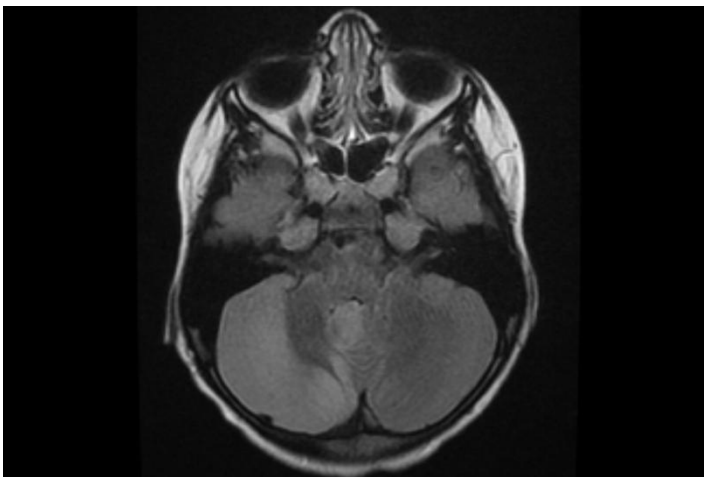

Fig. 1E. Axial T2 Flair with contrast

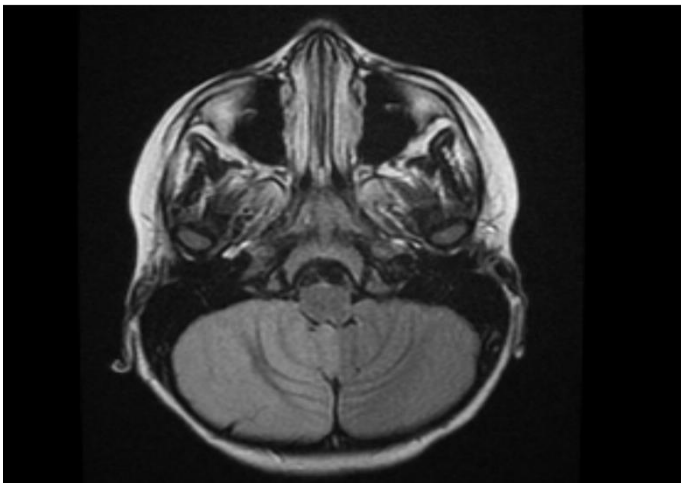

Fig. 1F. Axial T2 Flair with contrast

Fig. 1. Brain MRI: Unilateral right cerebellar abnormal FLAIR and T2 signal intensity with cortical swelling. No enhancement post-contrast administration. Most likely representing pseudo-tumoral hemi-cerebellitis

Virus, Human Adenovirus, Parainfluenza virus 1, Parainfluenza virus 2, Parainfluenza virus 3, Parainfluenza virus 4, Human Rhinovirus, Respiratory Syncytial Virus A, Respiratory Syncytial Virus B, Metapneumovirus, Human Coronavirus 229E, Human Coronavirus NL63, Human Coronavirus OC43, Human Bocavirus (HBoV), and Human Enterovirus (HEV).

After approximately 1 week the patient was discharged with significant improvement, and she was seen after 2 weeks in the clinic, the patient was completely back to her baseline with a normal neurological examination.

3. DISCUSSION

The patient had a unique presentation of the disease, in the form of unilateral weakness and areflexia. It had been postulated to divide the clinical presentation into two categories, the first one was cerebellar signs including ataxia, dysarthria, diplopia, nystagmus, intentional tremor, dysmetria and headache, and the second one was headache with other signs related to increased intracranial pressure but no cerebellar signs, the first group represented the majority in around 71.4% of the patients, the second group accounted for 28.6% [2]. However, the patient did not express headache, and initially, she had irritability, vomiting and fever which explained the infectious cause.

Pseudotumoral acute hemi-cerebellitis can be associated with the viral or bacterial infection in approximately 24% of the children and some patients had positive serological results for Epstein–Barr Virus, Influenza A virus, *Salmonella typhi*, Varicella Zoster Virus, and *Coxiella burnetii* [1,2]. In case of this patient, all viral studies as mentioned before were negative.

There were no clear signs of cerebellar involvement in the form of ataxia, dysarthria, diplopia, nystagmus, intentional tremor or dysmetria, in fact, she had unilateral weakness and areflexia. There was no clear reason for the weakness and not reported before as per the medical history.

The third unusual sign was the absence of both knee reflexes. In fact, the only role of cerebellum in the reflex arc had been that, just a few of the impulses from the spindle receptors through the arc after entering the dorsal horn would go to the cerebellum via the dorsal spinocerebellar tracts which play a critical role in cerebellar-cortical-spinal feedback loops to control balance and coordination which did not explain areflexia [9].

The patient had unilateral right cerebellar abnormal FLAIR and T2 signal intensity with cortical swelling and no enhancement. However, it was not one of the differentials because of the atypical presentation and the diagnosis was reached by MRI, that emphasised the role of MRI in such a patient.

Two studies explained the radiological findings in pseudotumoral hemi-cerebellitis patients [10,11] and vermis involvement has been described in (22.7%) of patients, all of them were younger than 15 years of age and all had unilateral cerebellar swelling, however, the patient did not showed any involvement.

For the treatment, it was reported that the steroid can be effective in shortening the duration of the disease, and in some patients, it can improve the long-term outcome (2). In case the patient had severe headache or disturbance of the level of consciousness together with cerebellar swelling on MRI steroid and mannitol use are both recommended, but if the patient showed severe obstructive hydrocephalus or signs of brainstem compression, the management would be surgical [1,3], however most of the patient did not require any treatment.

The prognosis was excellent but ipsilateral hemi-cerebellar atrophy may develop in up to 50% of cases, and a minority of them might have to persist fine motor and/or neurocognitive sequelae [1].

4. CONCLUSION

Pseudotumoral acute hemi-cerebellitis, an inflammatory condition, rarely reported, had a unique presentation in the form of headache, meningism and sometimes cerebellar signs. MRI was characteristic, which usually showed focal or diffuse signal changes in one cerebellar hemisphere with or without cerebellar swelling, the patient had a typical clinical picture in form of unilateral weakness and areflexia, which emphasised the importance for doing a neuro-image in such case, for diagnosis and then treatment purposes.

CONSENT

As per international standard or university standard, parent’s consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Lechón FC, Rodríguez AD, Cerdá SIS, Khabra K, de Prada I, García-Peñas JJ, et al. Hemicerebellitis: Report of three paediatric cases and review of the literature. European Journal of Paediatric Neurology. 2014;18(3):273-81.
2. Rodríguez-Cruz PM, Janet-Signoret S, Miranda-Herrero MC, Barredo-Valderrama E, Vázquez-López M, Ruiz-Martín Y, et al. Acute hemicerebellitis in children: Case report and review of literature. European Journal of Paediatric Neurology. 2013; 17(5):447-53.
3. Sawaishi Y, Takada G. Acute cerebellitis. The Cerebellum. 2002;1(3):223-8.
4. Horowitz MB, Pang D, Hirsch W. Acute cerebellitis: Case report and review. Pediatric Neurosurgery. 1991;17(3):142-5.
5. Jabbour P, Samaha E, Lahoud GA, Koussa S, Abadjian G, Nohra G, et al. Hemicerebellitis mimicking a tumour on MRI. Child's Nervous System. 2003;19(2): 122-5.
6. Levy EI, Harris AE, Omalu BI, Hamilton RL, Branstetter IV BF, Pollack IF. Sudden death from fulminant acute cerebellitis. Pediatric Neurosurgery. 2001;35(1):24-8.
7. Van Lierde A, Righini A, Tremolati E. Acute cerebellitis with tonsillar herniation and hydrocephalus in Epstein-Barr virus infection. European Journal of Pediatrics. 2004;163(11):689-91.
8. Desai J, Mitchell WG. Acute cerebellar ataxia, acute cerebellitis, and opsoclonus-myoclonus syndrome. Journal of Child Neurology. 2012;27(11):1482-8.
9. Lichstein PR. The medical interview; 1990.
10. De Bruecker Y, Claus F, Demaerel P, Ballaux F, Sciot R, Lagae L, et al. MRI findings in acute cerebellitis. European Radiology. 2004;14(8):1478-83.
11. Omeis IA, Khoshyomn S, Braff SP, Maugans TA. Idiopathic lymphocytic cerebellitis. Pediatric Neurosurgery. 2002; 36(1):52-3.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle3.com/review-history/48263