



Neuromelioidosis – A Misleading Clinical Presentation

Dhruvi Reddy Sanikommu ^{a*} and Murugesan Subramanian ^b

^a Department of Medicine, Mahatma Gandhi Medical College and Research Institute, Pondicherry, India.

^b Department of Neurology, Mahatma Gandhi Medical College and Research Institute, Pondicherry, India.

Authors' contributions

This work was carried out in collaboration between both authors. Both authors read and approved the final manuscript.

Article Information

DOI: 10.9734/AJMAH/2022/v20i530460

Open Peer Review History:

This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: <https://www.sdiarticle5.com/review-history/85638>

Case Study

Received 26 January 2022

Accepted 04 April 2022

Published 07 April 2022

ABSTRACT

This is a case of a young female who initially presented with visual symptoms, diagnosed as optic neuritis, and treated with steroids. She recovered completely with therapy and two days later, she developed left hemiplegia, dysarthria, and a drop in sensorium. Imaging with MRI brain was suggestive of demyelination changes with a possible diagnosis of primary CNS demyelination syndrome. With persistent fever spikes in-hospital and worsening sensorium, meningoencephalitis was also considered a differential. She was treated with empiric antibiotics and steroids. Blood culture grew burkholderia pseudomallaei, giving a definitive diagnosis of neuromelioidosis, a rare cause of meningoencephalitis. We thereby report this case to highlight the unusual presentation of neuromelioidosis masquerading as acute demyelinating encephalomyelitis, a challenging diagnosis for clinicians.

Keywords: Burkholderia; demyelination; meningoencephalitis.

1. INTRODUCTION

Neuromelioidosis is one of the rare neurological complications of burkholderia pseudomallaei

infection. Only around 1300 cases have been reported in the Asia-Pacific region since 2010. [1] It is endemic to areas in the Indian subcontinent, South East Asia, and Northern Australia. Very

*Corresponding author: E-mail: dhruvi.reddy24@gmail.com;

few cases have been reported in India so far. [2] It can present with features of encephalomyelitis on imaging, posing a diagnostic dilemma among physicians. This case focuses on the unusual clinical presentation and imaging findings of neuromelioidosis, the diagnosis of which was confirmed with blood culture.

2. CASE PRESENTATION

A 34 year old previously healthy South Indian woman presented to the outpatient clinic with complaints of impaired vision and blurring of vision in the left eye for one week. This was associated with intermittent headaches and giddiness. No other significant history.

On presentation, she was conscious and oriented. Vitals were stable. Ophthalmic examination revealed grade 2 relative afferent pupillary defect in the left eye. Neurological examination was otherwise remarkable. All routine investigations were within normal range.

Imaging with MRI brain showed symmetric T2 hyper-intensities in bilateral basal ganglia with optic nerve involvement, suggestive of possible demyelination. The patient was diagnosed as optic neuritis and was treated with pulse steroid therapy for 5 days. Her symptoms improved significantly after treatment.

However, she then started to develop fever spikes along with weakness of the left upper limb

and lower limb after the completion of therapy. This was associated with slurring of speech. No history of headache, giddiness, seizures, syncope or loss of consciousness. No other relevant history was noted.

On this presentation, the patient was drowsy but was obeying commands. Vitals – Temperature 99 degrees Fahrenheit, blood pressure 110/70 mmHg, pulse rate 92 beats per min. Neurological examination revealed slurring of speech, decreased tone in left upper and lower limb with power of 0/5 and plantar extensor. Routine investigations revealed elevated blood counts with neutrophilic predominance, elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), mild transaminitis, and normal renal parameters.

MRI brain showed patchy enhancement at the right pre-central gyrus and small central nodular enhancing areas within and adjacent subcortical white matter with surrounding excessive perilesional white matter edema [Fig. 1], also extending along the right corticospinal tract up to midbrain level [Fig. 2]. Similar patchy enhancement with small nodular enhancement was noted within at left medial lentiform nucleus on inferior aspect with surrounding edema [Fig. 3]. Small ring-enhancing lesions were also noted at the right hippocampal gyrus with subtle perilesional edema.

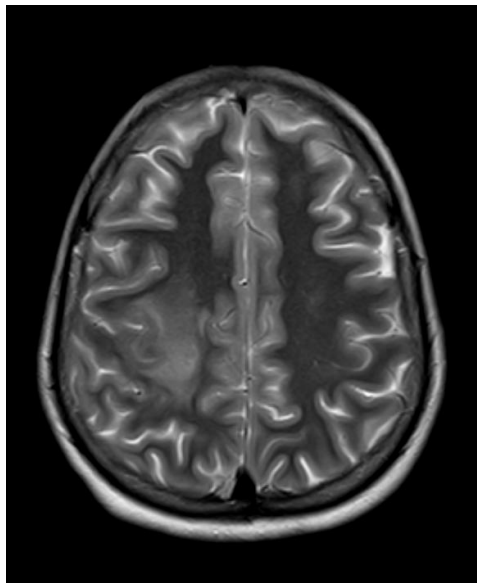


Fig. 1. MRI brain showing patchy enhancement of precentral gyrus with perilesional white matter edema

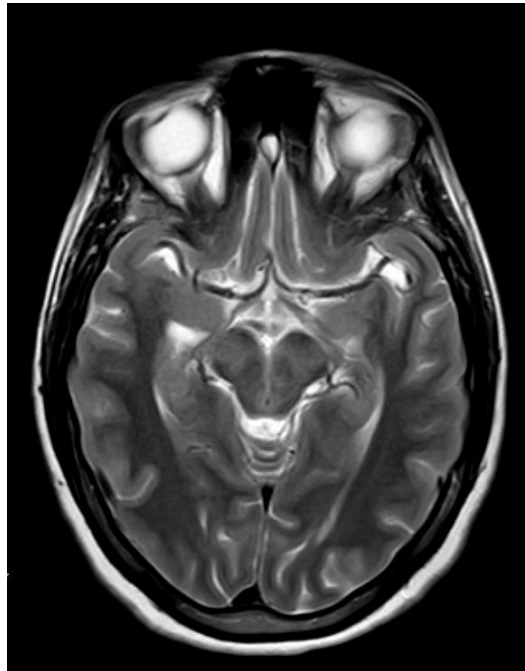


Fig. 2. MRI brain showing perilesional edema further extending up to midbrain

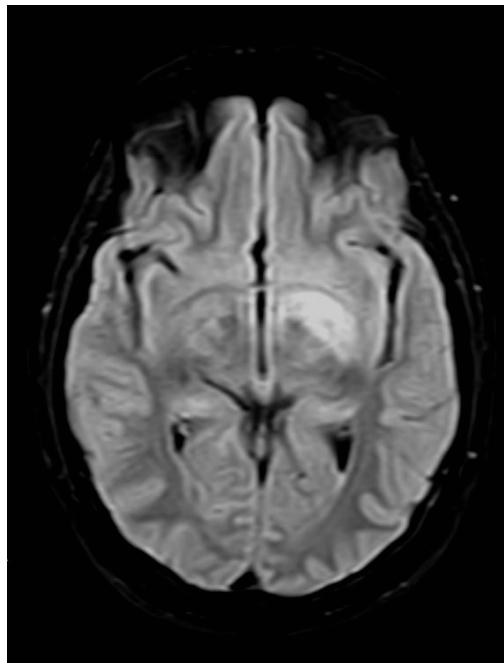


Fig. 3 Shows patchy enhancement in left medial lentiform nucleus with surrounding edema

During the hospital stay, patient developed persistent fever spikes and fall in sensorium. The differentials thought of were primary CNS demyelination syndrome and meningoencephalitis. She was treated with empiric antibiotics and steroids. Serology for HIV was negative. Cerebrospinal fluid (CSF) analysis showed elevated counts of 430 cells/mm³ with

lymphocyte predominance (80%) along with low glucose and normal protein levels. CSF gram stain revealed occasional pus cells, no organisms. CSF Adenosine deaminase was normal and culture was sterile. However, blood culture grew *Burkholderia pseudomallei*. [Figs 4, 5 and 6].



Fig. 4. Gram stain showing gram negative bacilli with safety pin appearance



Fig. 5. Blood agar showing glistening grey white colonies



Fig. 6. MacConkey agar showing pale pink colonies

Antibiotics were changed to ceftazidime and cotrimoxazole according to culture sensitivity. The patient developed a worsening sensorium and was electively intubated. Antibiotics were continued and fever spikes subsided. Due to patient's financial constraints, she was taken to hospital of choice. On telephonic follow-up, it was brought to our notice that the patient succumbed to death one week after discharge.

3. DISCUSSION

Melioidosis is caused by a gram negative, facultative soil saprophyte which is acquired via wet soil and water inoculation through wounds or inhalation. [3] It has a varied presentation, mainly involving sites of pulmonary, followed by genitourinary, and skin. [4] Neuromelioidosis is one of the rare neurological complications of burkholderia pseudomallei infection, which accounts for approximately 4% of all melioidosis cases [5]

This infection can be latent for prolonged periods leading to further reactivation of the disease later, like in CNS tuberculosis. Hematogenous spread of the microorganism to the central nervous system plays a key role in disease pathogenesis. [4] This is supported by this study's result that blood is the most common specimen that isolated *B. pseudomallei*. The circulating bacteria can cross the cellular barriers, which include the blood-brain barrier and blood-CSF barrier, using various methods: direct brainstem invasion, percutaneous inoculation, transcellular, paracellular, or Trojan horse method. [3,4]

Clinical manifestations of neuromelioidosis vary from encephalomyelitis to seizures and focal neurological deficits with cranial nerve involvement. Fever is the most common presenting feature (74%). [3,4]

On imaging with MRI brain, isolated supratentorial lesions were found to be the most commonly involved structure in CNS melioidosis. [4,6] Patients can also have rim-enhancing micro abscesses with propensity for white matter tracts including the corticospinal tract, corpus callosum and cerebellar peduncles. [7]

This patient initially presented with optic neuritis and symptomatically improved with steroid therapy. However, she developed progressive neurological deficits of hemiplegia with dysarthria, suspecting acute demyelinating

encephalomyelitis (ADEM) given both clinical and imaging findings. Due to persistent fever spikes and non-improvement of clinical condition with steroids, an infective cause was thought to be more likely. There was a diagnostic dilemma between ADEM and meningoencephalitis. CSF findings of high counts with lymphocyte predominance and normal glucose and proteins levels however did not support the diagnosis of ADEM in this patient.

In ADEM, MRI brain demonstrates multiple hyperintense bilateral, asymmetric patchy lesions, involving subcortical and central white matter, cortical gray-white matter junction, thalami, basal ganglia, and brainstem. [8] In this patient, the first MRI brain showed hyperintensities in bilateral basal ganglia with optic nerve involvement suggestive of a demyelinating pathology. However, repeat imaging revealed both supra- and infratentorial structure involvement along with excessive white matter edema extending into the corticospinal tract upto midbrain level. Small ring enhancing lesions were also noted. This invasive spread is similar to conditions like fungal infections and tuberculosis. Neuromelioidosis is a great mimicker of tuberculosis, with ring-enhancing lesions on brain MRI. Lesion progression with involvement of the corticospinal tract may be pathognomonic for this condition. [9] The diagnosis was hence confirmed with blood culture growing burkholderia pseudomallei.

In neuromelioidosis, ceftazidime and meropenem are the drugs of choice for intensive-phase therapy according to the 2010 consensus recommendations. Cotrimoxazole is the drug of choice for eradication-phase therapy. [10] This patient was treated with ceftazidime and cotrimoxazole according to culture sensitivity. There was an improvement in blood counts and a reduction in fever spikes. However, rapid deterioration of the patient's sensorium was the cause of mortality in this patient. Melioidosis affecting CNS has a low incidence but significantly high morbidity and mortality. [11,12]

4. CONCLUSION

Neuromelioidosis can masquerade as acute demyelinating encephalomyelitis, and pose as a challenging diagnosis for clinicians. Its atypical presentation must be aware to all and should be suspected in nonendemic areas as well. Considering high mortality rates, early diagnosis and initiation of antibiotic therapy are of utmost importance.

ETHICAL APPROVAL

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

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Zamzuri MAIA, Jamhari MN, Nawi HM, Hassan MR, Pang NTP, Kassim MAM, Rahim SSSA, Jeffree MS, Lee SY. Epidemiology of Neuromelioidosis in Asia-Pacific: A Systematic Review. Open Access Macedonian Journal of Medical Sciences. 2021;9(F):318-326
2. Ekka A, Mohideen M, Kesavan S. Neuromelioidosis masquerading as acute demyelinating encephalomyelitis. Indian Pediatrics. 2017;54(12).
3. Nagendra S, Shah AG. Neuromelioidosis—A tropical illness presenting as meningoencephalitis with unusual brain imaging. Annals of Indian Academy of Neurology. 2021;24(1):97.
4. Wongwandee M, Linasmita P. Central nervous system melioidosis: a systematic review of individual participant data of case reports and case series. PLoS neglected tropical diseases. 2019;13(4):e0007320.
5. Arif MA, Abid MH, Renganathan R, Siddiqui KA. Central and peripheral nervous system involvement in neuromelioidosis. BMJ Case Reports, bcr. 2015;211001.
6. Hsu CC, Singh D, Kwan G, Deuble M, Aquilina C, Korah I, Norton R. Neuromelioidosis: craniospinal MRI findings in Burkholderia pseudomallei infection. Journal of Neuroimaging. 2016; 26(1):75-82.
7. Pohl D, Alper G, Van Haren K, Kornberg AJ, Lucchinetti CF, Tenembaum S, Belman AL. Acute disseminated encephalomyelitis: updates on an inflammatory CNS syndrome. Neurology. 2016;87(9 Supplement 2):S38-45.
8. Lipsitz R, Garges S, Aurigemma R, Baccam P, Blaney DD, Cheng AC, et al. Workshop on treatment of and postexposure prophylaxis for Burkholderia pseudomallei and B. mallei infection, 2010. Emerg Infect Dis. 2012;18:e2.
9. Zhan Y, Wu Y, Li Q, Yu A. Neuromelioidosis: a series of seven cases in Hainan province, China. Journal of International Medical Research. 2017; 45(2):856-67.
10. Mannam P, Arvind VH, Koshy M, Varghese GM, Alexander M, Elizabeth SM. Neuromelioidosis: A Single-Center Experience with Emphasis on Imaging. Indian Journal of Radiology and Imaging. 2021;31(01):057-64.
11. Chatterjee A, Saravu K, Mukhopadhyay C, Chandran V. Neurological Melioidosis Presenting as Rhombencephalitis, Optic Neuritis, and Scalp Abscess with Meningitis: A Case Series from Southern India. Neurol India. 2021;69:480-2.
12. Jabeen S, Saini J, Mishra T, Mailankody P, Lingaraju TS, Chandrashekar N. Neuromelioidosis presenting as stroke like syndrome. Neurology: Clinical Practice; 2020.

© 2022 Sanikommu and Subramanian; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:

The peer review history for this paper can be accessed here:
<https://www.sdiarticle5.com/review-history/85638>