An unusual case of Acute Disseminated Encephalomyelitis (ADEM) and the diagnostic challenges involved in managing patients with Human Immunodeficiency Virus.

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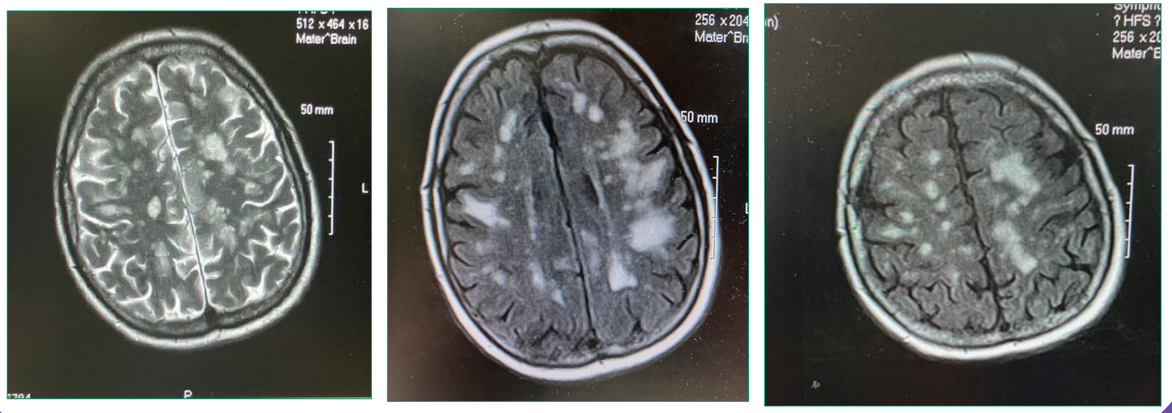
Background & Methods

- People living with HIV (PWH) are at increased risk of neurological comorbidities compared to the general population despite treatment with effective antiretroviral treatment (ART).
- We present a case highlighting the diagnostic challenges arising from a rare debilitating neurological condition in an individual with HIV.

Case History

- A 67-year-old man living with HIV for 17 years on effective ART but with sub-optimal immunological response (CD4+ cell count <200 copies/ml) presented to a Dublin Hospital with abdominal pain, constipation and urinary retention.
- Past medical history:
  - Diffuse Large B-cell lymphoma (2018)
  - Idiopathic thrombocytopenic purpura. , and despite negative testing on CSF, empiric treatment for neurosyphilis was completed with little response
- Initial CT Brain was normal, however over 3 days he became encephalopathic and aggressive, dropping his GCS, requiring intubation and transfer to critical care in the Mater Hospital.

MRI Brain



Investigations

- MRI showed extensive, non-enhancing T2 hyperintense foci within the cerebral white matter, brain stem, and right cerebellar hemisphere in addition to multiple foci in the cervical and thoracic spinal cord.
- Lumbar puncture had normal opening pressure, cell count and biochemical parameters.
- Investigations for an infective cause were unremarkable:
  - Normal serum and CSF cryptococcal antigen
  - CSF HIV, JC virus and Toxoplasma PCR: Negative
  - CSF geneXpert for MTB and TB culture: Negative
  - Broad range 16s and 18s PCR.
  - Treponema Pallidum serology was positive with a known history of treated infection.
- CT thorax, abdomen and pelvis were unremarkable with repeated large volume.
- CSF cytometry showed no evidence of lymphoma recurrence.

Working diagnosis and Management

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- Palladium serology was positive with a known history of treated infection, and despite negative testing on CSF, empiric treatment for neurosyphilis was completed with little response
- Following the above investigations and in consultation with Neurology colleagues a final working diagnosis of Acute disseminated encephalomyelitis(ADEM) was reached.
- Following courses of intravenous immunoglobulin and high dose dexamethasone, there was significant clinical improvement allowing extubation followed by improvement in GCS and ICU discharge after 21 days.
- Repeat MRI showed significant improvement in both the size and intensity of the white matter foci. He subsequently underwent inpatient rehabilitation.

Conclusion

- ADEM is a rare immune-mediated inflammatory, demyelinating CNS disorder, commonly preceded by infection(1).
- Although more common in children, ADEM occurs in adults and has been described in HIV infection often presenting in an atypical fashion(2).
- Given the broad differential of CNS pathology in immunosuppressed PWH and the atypical features of ADEM in this population, the diagnosis of ADEM can be challenging and is often delayed.
- Clinicians must maintain a broad diagnostic differential when PWH present with neurological conditions.