

Acute Disseminated Encephalomyelitis following Thoracic Endovascular Aortic Repair: An unusual presentation

Dr. Cleofina Furtado (✉ cleofinafurtado@gmail.com)

University Hospitals of North Midlands <https://orcid.org/0000-0003-3726-0837>

Dr. Changrez Jadun

University Hospitals of North Midlands

Dr. Sachin Srivastava

University Hospitals of North Midlands

Dr. Sanjeev Nayak

University Hospitals of North Midlands

Dr. Zafar Hashim

University Hospitals of North Midlands

Case Report

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Abstract

Acute disseminated encephalomyelitis (ADEM) is a central nervous system demyelinating condition caused by viral or bacterial infections or immunizations. The postulated aetiology is an autoimmune reaction against myelin components; however, the mechanistic details are yet unknown. We present the imaging findings of a patient who passed away due to the manifestations of acute disseminated encephalomyelitis. The patient initially underwent surgical repair for type B aortic dissection. 2 weeks after he had poor GCS and no motor response following multiple attempts at sedation hold in the intensive care unit. On magnetic resonance imaging, haemorrhagic white matter abnormalities were noted throughout the cerebral hemisphere indicative of an inflammatory demyelinating process.

Our case emphasizes the challenges involved in evaluating emerging neurological problems following a period of intubation /sedation amongst these patients. In addition, unlike children, 50% of adult patients do not have illness or immunisation as a trigger event for this condition.

Background

Acute Disseminated Encephalomyelitis (ADEM) is a virus or vaccine-induced monophasic inflammatory myelinating autoimmune reaction against myelin (1). ADEM often occurs relatively late after the beginning of infection and is distinguished by increased white matter damage (2). Encephalopathy with rapid neurologic impairments and brain magnetic resonance imaging (MRI) findings consistent with multifocal demyelination describe this condition (3). It is more common in children and young adults, while occurrence in middle age or elderly individuals are uncommon (4,5).

SARS-CoV-2, varicella-zoster, Measles, rubella, mumps, Epstein Barr virus, coxsackievirus, cytomegalovirus, hepatitis A, and herpes simplex virus are all viral diseases linked to ADEM (6–9). Mycoplasma pneumonia is the most common, with leptospira, Borrelia borgdorgeri, and group-A beta-hemolytic streptococci being the other bacterial illnesses (7,9). Anti-rabies, Pertussis, diphtheria, measles, mumps, rubella, and influenza immunizations are also linked to ADEM with growing cases also noted post-COVID-19 vaccination in the recent era (9,10).

We present this unique case of ADEM which occurred following thoracic endovascular aortic repair. Because of its unique occurrence, presentation of the disease and prognosis, it was found worthy to be mentioned.

Case Description

A 62-year-old patient was hospitalised in the intensive care unit following an elective thoracic endovascular aortic repair of type B aortic dissection. He was intubated and ventilated subsequently due to worsening breathing and requirement for oxygen. Chest radiographs (Fig 1 a) and computed tomography did not reveal any untoward. There was no history of immunization, infectious or vaccination in recent days. He had no known chronic illness or drug use. Magnetic resonance imaging (MRI) was performed, as he had poor GCS and no motor response following multiple attempts at sedation hold with no recovery of consciousness which was 2 weeks following the TEVAR. It showed multiple patchy confluent T2WI and FLAIR hyperintense lesions (Fig 2) involving cerebral white matter (callosal, pericallosal, deep, periventricular, basal ganglia), brainstem and bilateral cerebellar peduncles. These lesions demonstrated diffusion restriction (Fig 3) with some showing avid contrast enhancement. Susceptibility weighted imaging sequence (Fig 4) demonstrated multiple foci of hemosiderin deposition in the cerebral cortices bilaterally, thalamus and cerebellar hemispheres. Findings were concluded to be characteristic of demyelination such as acute disseminated encephalomyelitis (ADEM). Lumbar puncture revealed lymphocytic pleocytosis with slightly raised protein and absence of oligoclonal bands. The EEG demonstrated generalised slowing of sleep activity.

He also developed multi-organ failure with anuria requiring continuous hemodiafiltration. Over the next few days, he remained critically unwell with a high level of inflammatory markers and no signs of neurological improvement. Following a discussion with his family, it was decided to withdraw support following which he passed away.

Discussion

TEVAR has enabled a less invasive method to treat a variety of thoracic aortic diseases and is generally performed in surgically repaired type A dissection and those with a complicated type B dissection (11). ADEM after TEVAR has not yet been documented. Radiopaque contrast medium might have triggered an immune-mediated response, resulting in ADEM.

ADEM is known to occur as a result of an inflammatory response to a foreign antigen that causes vascular congestion and increased permeability of the vasculature of the central nervous system (12). This is expected to set off an inflammatory cascade including oedema, inflammatory cell infiltration and perivenous haemorrhage leading to gliosis, demyelination and necrosis (13).

Neurologic symptoms are variable, with the majority exhibiting encephalopathy, which includes confusion, and lethargy, with difficulty awakening from sedation as the most common presentation (14). These characteristics have the potential to hide an underlying inflammatory central nervous system

condition in the critical care setting, where clinical neurological assessment is particularly difficult (6). In addition, para infectious neuropathological processes often manifest after a latent time following an infection, raising the clinical challenge of when to evaluate emerging neurological problems following a period of intubation along with challenges involved in transporting the patient for more complex imaging studies (15).

Histology is characterised by tiny (5mm) white matter lesions with macrophage clusters and a variety of related axonal damage (15). The widespread haemorrhagic white matter lesions identified have some of the characteristic features of acute haemorrhagic necrotizing encephalitis which is known to have a fulminant (16) .

MRI imaging lesions show lesions with high sensitivity and may help rule out other entities that look similar, such as multiple sclerosis, which displays lesions around the deep white matter, near the corpus callosum or calloso septal junction (17). FLAIR hyper-intensities in deep white matter and at the grey/white matter interface with diffusion restriction are typical MRI lesions. Punctate or rim enhancing lesions are seen on post-contrast enhancement (18).

Conclusion

Whilst infection and vaccination are the most common associations for ADEM, we suspect that radiopaque contrast material might have been a possible immunological trigger element in this case. The patient's clinical course demonstrates that during lengthy hospitalizations, neurological manifestations of ADEM might arise which are especially difficult to assess and recognize in critically unwell patients.

Declarations

Conflict of interest:

None declared.

I/We confirm that written informed consent for the case to be published (including radiological images and case history) was obtained from the patient's next of kin for publication of this case report.

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Figures

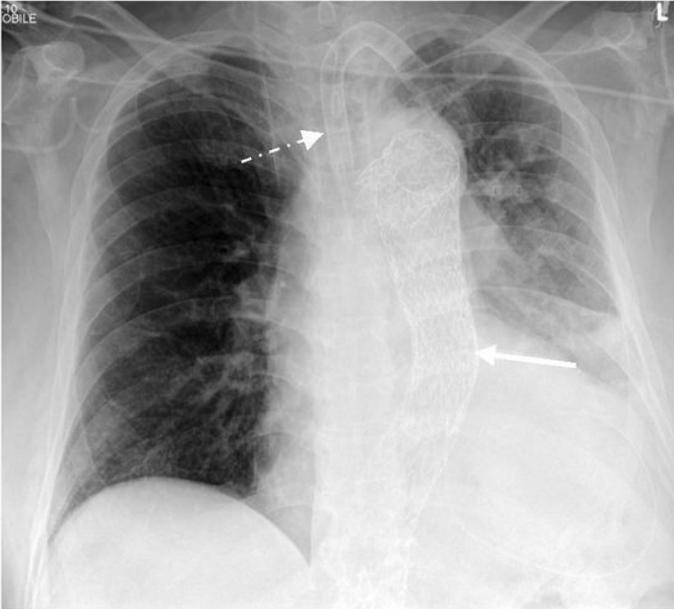


Fig 1

Supine AP chest radiograph

Note post-TEVAR metallic stent (white solid arrows) and ET tube (white dashed arrow).

Figure 1

See image above for figure legend.

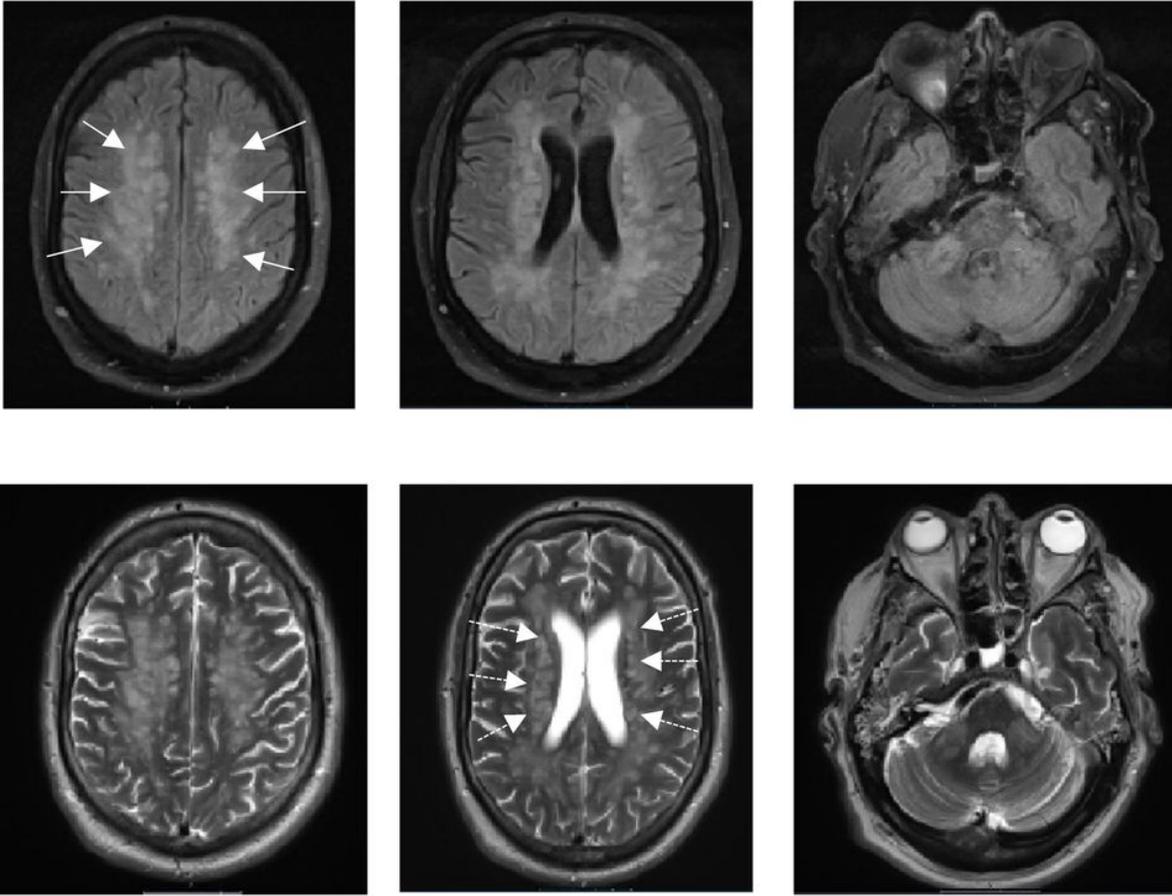


Fig 2

MRI: Axial FLAIR (top row) and Axial T2 (bottom row) demonstrate periventricular white matter high signal intensities on FLAIR (white solid arrows) and T2WI (white dashed arrows).

Figure 2

See image above for figure legend.

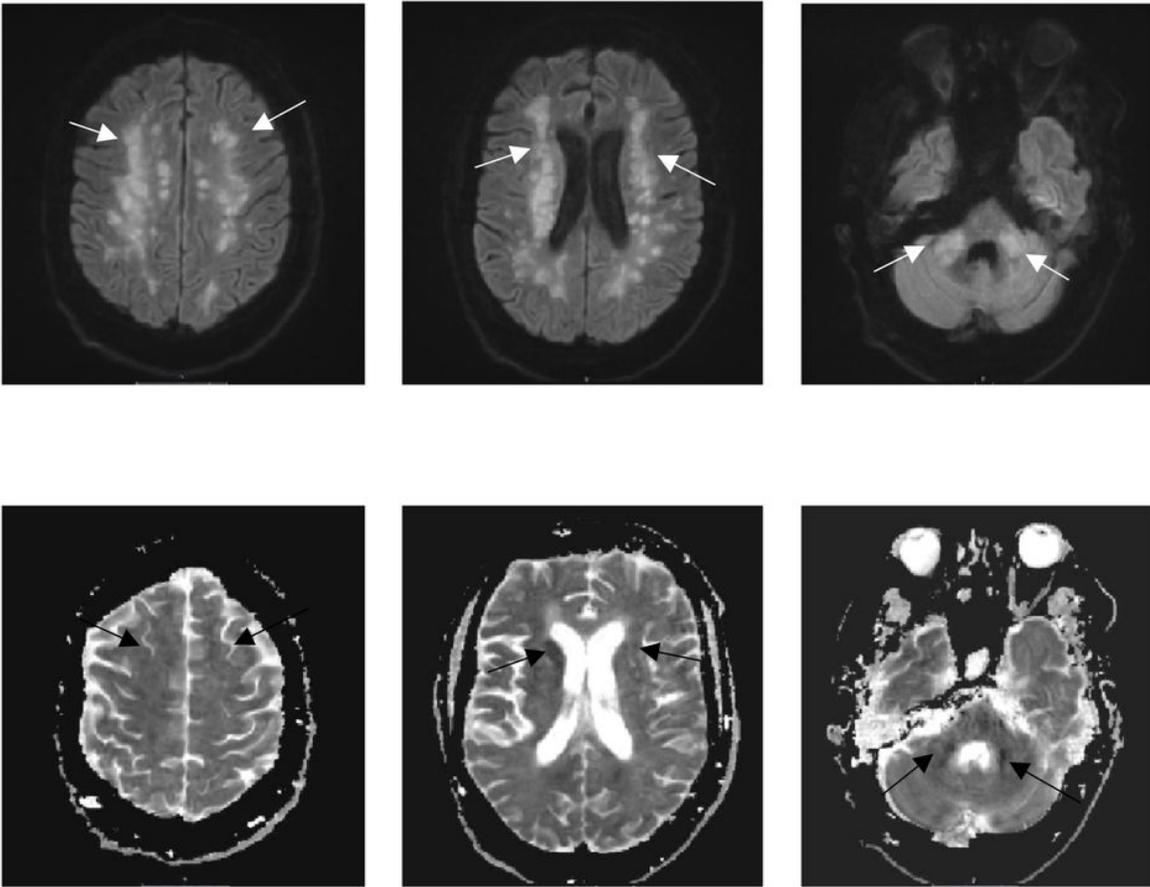


Fig 3

MRI: Axial DWI (top row) and ADC (bottom row) demonstrate abnormal diffusion restriction with high signal on DWI (white arrows) and low signal on ADC map (black arrows) in the corresponding white matter lesion.

Figure 3

See image above for figure legend.

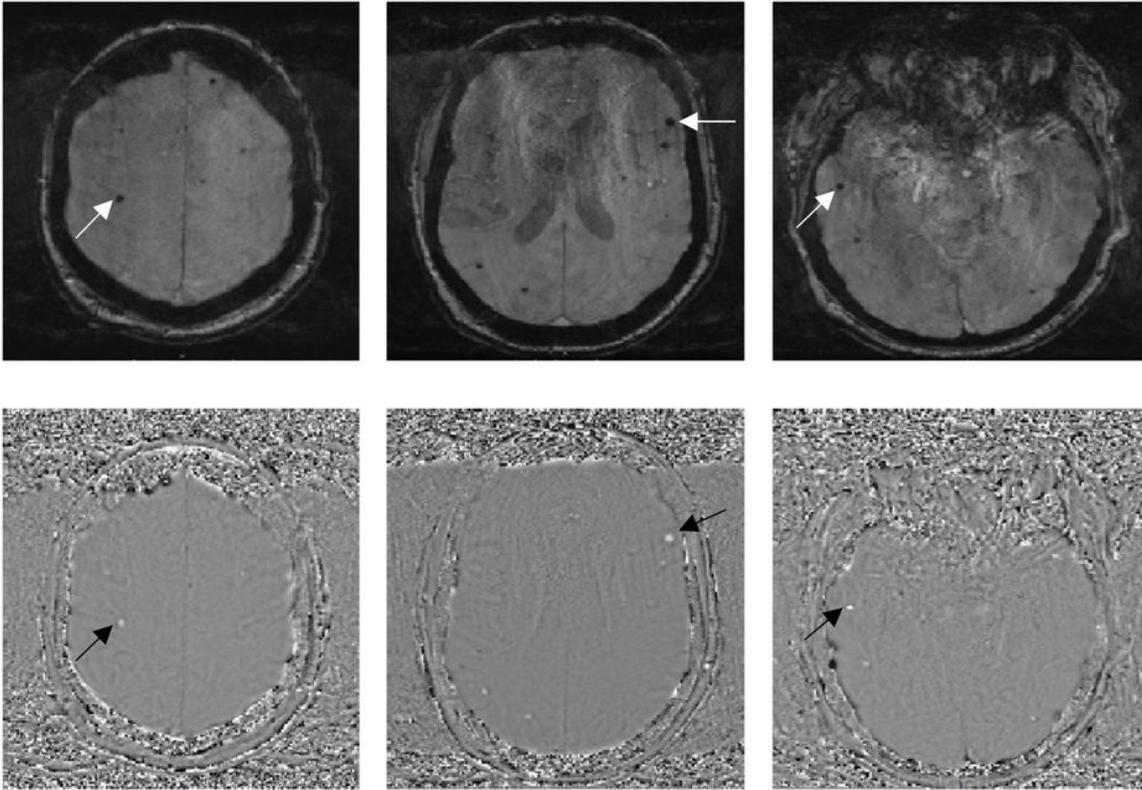


Fig 4

MRI: Axial susceptibility weighted imaging (top row) demonstrates tiny areas of blooming artifacts (white arrows). Corresponding phase imaging (bottom row) shows high signal (black arrows) keeping with hemosiderin deposition.

Figure 4

See image above for figure legend.