Hepatitis E-associated neuralgic amyotrophy: a rare respiratory presentation

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Introduction

Neuralgic amyotrophy is characterised by acute neuropathic pain, motor dysfunction with or without sensory loss. Most cases present with these classical or typical core symptoms. A 55-year-old male presented with no core symptoms, but with flu-like symptoms and central chest pain. A medical history of hepatitis E virus infection 6 months before the current presentation and his present symptoms led to a clinical suspicion of neuralgic amyotrophy, which was confirmed by diaphragmatic weakness and absence of any other definite aetiology. Neuralgic amyotrophy can be difficult to diagnose, but an awareness of atypical presentations and hepatitis E virus association can address patient concerns, target management and avoid unnecessary investigations.

Case report

A 55-year-old man, with no significant past medical history, initially presented with jaundice, feeling generally unwell and lethargy. He was investigated in the infectious diseases' unit for deranged liver function tests and this confirmed hepatitis E (confirmed with IgM and IgG serology for hepatitis E virus infection). The rest of the liver screen bloods were unremarkable, including those for hepatitis A, B and C, human immunodeficiency virus (HIV), antinuclear antibodies, antimitochondrial antibodies, anti-smooth muscle antibodies and anti-liver-kidney-microsome antibodies. Ultrasound of his liver demonstrated diffuse fatty liver changes with otherwise no evidence of focal lesion or dilated ducts. Leptospirosis serology was sent considering his work in a water treatment plant and potential exposure to sewage; this was later reported negative.

The patient presented 6 months later with flu-like symptoms and a sharp acute central chest pain accompanied by dyspnoea. Chest X-ray (Figure 1b) revealed a raised left hemidiaphragm; an abnormality confirmed by computed tomography (Figure 2). No additional abnormality was found on either radiological investigation. There was no clinical evidence of acute infection during his second presentation and his blood tests and arterial blood gases were normal. He was observed overnight and did not require additional investigations as an inpatient. He had received the yearly flu vaccine 8 months before his initial presentation and 2 months before his second.

Diaphragmatic ultrasound confirmed reduced movement, with respiratory muscle assessment demonstrating a reduction in inspiratory pressure. A few weeks after the second presentation, the patient's chest pain resolved and his breathlessness improved over time but not back to his baseline.

Given the suspicion of hepatitis E virus-induced neuralgic amyotrophy, overnight oximetry, nerve conduction studies and respiratory muscle assessment were arranged to both confirm and determine the extent of neuralgic amyotrophy. The second presentation was deemed to be a sequela of his initial presentation which led to further investigations and confirmed respiratory muscle weakness secondary to neuralgic amyotrophy.

The diagnosis was supported by the following investigations:

- IgM and IgG serology for hepatitis E virus infection
- Chest X-ray and computed tomography of the thorax confirmed the elevated left hemidiaphragm position (Figure 1b and Figure 2)
- Thoracic ultrasound of the chest confirmed limited movement of the diaphragm
- Magnetic resonance imaging of the neck ruled out any cervical spine or other pathology leading to weakness
- Sniff nasal inspiratory pressure showed reduced diaphragmatic strength.

How to cite this article:
Basit RH, Ali A, Mustfa N, Thomas A. Hepatitis E-associated neuralgic amyotrophy: a rare respiratory presentation. Br J Hosp Med. 2023. https://doi.org/10.12968/hmed.2022.0450

Case report (continued)

His overnight oximetry showed few rapid eye movement sleep-related desaturations but was otherwise normal. As his symptoms were improving, a watch and wait approach was adopted and he did not require any intervention for his respiratory muscle weakness. A follow up in 6 months showed improving lung function and corresponding improvement in his radiology (Figure 1c). His sniff nasal inspiratory pressure improved from -42 to -60 cmH_2O and his forced vital capacity improved from 4.07 litres to 4.64 litres.

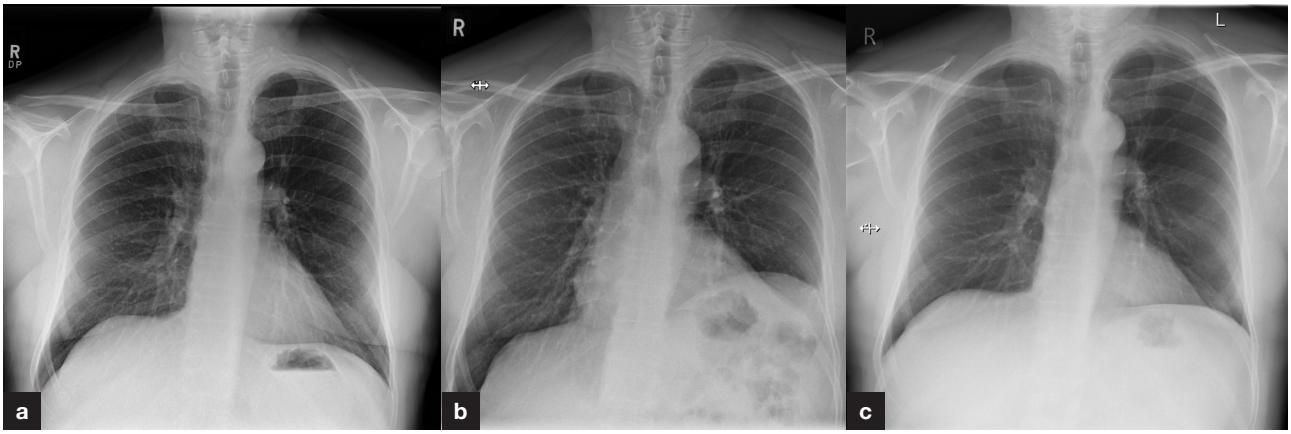

Figure 1. a. Chest X-ray on initial presentation. b. Chest X-ray showing raised left hemidiaphragm 6 months later. c. Chest X-ray with recovery of left hemidiaphragm 12 months after initial presentation.

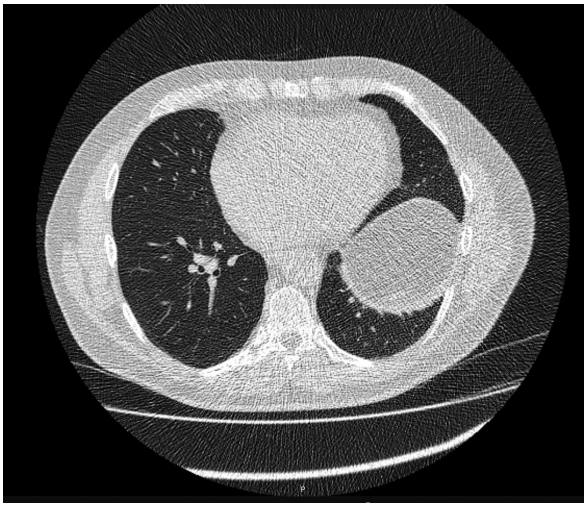

Figure 2. Computed tomography scan 6 months after initial presentation confirming the left hemidiaphragm.

Discussion

Neuralgic amyotrophy is more common in men than women, and almost always presents asymmetrically (van Eijk et al, 2014). The classical pattern of neuralgic amyotrophy typically involves a painful phase characterised by acute onset neuropathic pain, followed by unifocal or multifocal paresis (traditionally in the upper limb) and atrophy, followed by sensory disturbances before recovery (Jahic et al, 2019). This patient presented with flu-like symptoms, dyspnoea and central chest pain, leading clinicians to consider respiratory and/or cardiac pathologies. The only symptom potentially associated with neuralgic amyotrophy was central chest pain, but even this was atypical as over 95% of patients with neuralgic amyotrophy complain of pain in the neck, shoulder and/or arms. The absence of significant pathology on further radiological and physiological assessment and past medical history of hepatitis E virus infection prompted targeted investigations for neuralgic amyotrophy.

Learning points

- Atypical presentations of neuralgic amyotrophy need to be considered.
- Hepatitis E infection is associated with neuralgic amyotrophy.
- The management of neuralgic amyotrophy is largely supportive and is best managed by the speciality in which the symptoms predominate.

Previous studies have associated hepatitis E virus infection with neuralgic amyotrophy and thus the authors consider it vital for clinicians to consider this diagnosis in patients with previous hepatitis E infection, albeit without the classical neuralgic amyotrophy symptom triad (Theochari et al, 2015). There is no literature on the standard timeframe between hepatitis E infection and the development of neuralgic amyotrophy. The typical presentation period between viral infections and other post-viral neurological manifestations is about 24 days, up to 1–2 months (Filosto et al, 2021).

Neuralgic amyotrophy has a low incidence of 2 in 100 000 patients, but this is likely to be an underestimate as a community-based prospective study demonstrated that classical neuralgic amyotrophy was at least 30–50 times more common than currently accepted, finding an incidence rate of 1 in 1000 (van Alfen et al, 2015). They attributed this discrepancy to a lack of awareness of neuralgic amyotrophy, with which the authors agree.

The authors consider it vital that clinicians explore associations such as previous hepatitis E virus infection in atypical cases of neuralgic amyotrophy to increase diagnostic sensitivity. This will help address patient concerns, target management and avoid unnecessary investigations.

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