Acute Dysphagia as the Presenting Feature of Syringobulbia in a Sixteen-Year-Old Patient

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Introduction

Doctors often use heuristics (pattern recognition) to aid in the diagnostic process. This case emphasises the importance of keeping an open mind about the pathology behind a presenting complaint and the relevance of multi-speciality cooperation.

Case History

16-year-old female, with no significant past medical or drug history, presented with “worsening sore throat, cough, unable to swallow + drooling of saliva, feeling increasingly unwell” [A&E notes]. She reported:

- 1-week history of increasing dizziness, nausea and dry wretching episodes
- 2-week history of bowels not opening
- 3-week history of fevers and increased amount of phlegm
- Reduced oral intake due to inability to swallow (painless) “it doesn’t go any further”

She lives with her family (both teachers) and recently had a GCSE Maths exam.

The child was admitted to hospital.

Timeline

6th Jan 2015
05:39
Admitted under the medical team.
Neurological examination: Normal eye movements, normal power and sensation throughout lower limbs.
11:42
Post-Take Ward Round: History and bloods reviewed (Fig. 1).

Neurological examination: Downgoing plantars.

Impression: Need to rule out meningitis.

Initial management: Intravenous Aciclovir & Ceftriaxone, CT Head, lumbar puncture after CT Head.

12:04
CT Head: Medulla and cervical cord are expanded by a centrally-located longitudinal cerebrospinal fluid-density lesion.

References


Discussion

• Haemangioblastoma are vascular tumours in the spinal cord which can cause Syringomyelia. They are uncommon tumours of the Central Nervous System (CNS), accounting for less than 3% of all CNS tumours. They are usually well-circumscribed, highly-vascular and benign.

• Syringomyelia can present with a protean range of neurological deficits. Case reports in literature show that dysaesthesiae, quadriparesis, hiccups, headaches, ataxia and cerebellar signs could all be presenting signs of Syringomyelia or Haemangioblastoma. They can also exist in asymptomatic patients.

• If Syringomyelia is still suspected after CT imaging of the CNS, regular neurological examination should be carried out to detect evolving neurological signs and prompt referral to Neurosurgery is warranted.

• Von Hippel Lindau Disease (VHL) is an autosomal dominant condition characterised by haemangioblastoma in the CNS, retina, kidneys, adrenal glands, endolymphatic sac and pancreas. A 2014 analysis of 92 cases of patients who underwent resection of intraspinal haemangioblastoma showed that 35% had VHL.

Learning Points

Think of Bulbar/Pseudobulbar palsies in any patients with dysphagia or has neurological deficit – including the young.

Neurological signs can change quickly – regular full neurological examination is key!