Case Report

CNS infection with a history of recurrent epistaxis: Streptococcal meningitis as a first presentation of juvenile nasopharyngeal angiofibroma

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Abstract

An adolescent male with a history of recurrent epistaxis presented with headache and vomiting. Investigations revealed concurrent meningitis as well as the presence of a subarachnoid haemorrhage. Subsequent imaging identified a Juvenile Nasopharyngeal Angiofibroma; a rare but important cause of meningitis that should be considered in the young adult population.

Keywords

Meningitis, Juvenile Nasopharyngeal Angiofibroma, Epistaxis

Learning Points

1. Meningitis requires prompt treatment but also consideration of an underlying aetiology.
2. Although typically a paediatric disease, the young adult population affected can easily present to the adult acute take.
3. A history of epistaxis or rhinorrhea in an adolescent male presenting with meningitis should prompt further imaging and concern for Juvenile Nasopharyngeal Angiofibroma.

Summary

We present the case of a 17-year-old male developing streptococcal meningitis complicated by apparent subarachnoid haemorrhage, with an extremely rare underlying aetiology. The diagnosis of Juvenile Nasopharyngeal Angiofibroma is a rare but important one. Although often considered a paediatric pathology, the adolescent patient will often, as in this case, present to adult medicine when 16 years or over. We suggest that young males presenting with meningitis should be considered for CT and MR imaging if there is history of epistaxis, nasal obstruction, or rhinorrhoea.

Case Presentation

A 17 year old male presented with four days of headache with associated nausea and vomiting. He was normally fit and well with no regular medications. Of note he reported frequent epistaxis in the weeks prior to presentation and for several years had suffered from at least monthly epistaxis. Examination revealed pyrexia of 38.3, positive kernig's sign and photophobia inhibiting fundoscopy. There was no rash, the patient was haemodynamically stable and his Glasgow Coma Scale (GCS) was 15. Initial investigations identified raised inflammatory markers; WCC 13.2 x 10^9/L (4-11 x 10^9/L) and CRP 102 mg/L (0 -10 mg/L) with a normal chest X-ray and electrocardiogram.

He was diagnosed provisionally as bacterial meningitis and treated with IV ceftriaxone and oral dexamethasone. In the absence of signs of raised intra cranial pressure, a lumbar puncture (LP) was performed. Opening pressure was significantly raised at 40mmH20 (6-20cmH20) and the cerebrospinal fluid (CSF) was noted to be cloudy. CSF analysis revealed WCC > 3000x10^6/L (0-5 x 10^6/L) with 75% neutrophils; RBC 640x10^6/L (0-10 x 10^6/L); glucose < 0.6 mmol/L and a protein of 7.3 g/L (0.15-0.4 g/L). Cultures grew *Streptococcus Pneumococci*. Interestingly, xanthochromia was also present and a CT head was completed. The CT head identified blood in the lateral ventricles, right sylvian fissure and suprasellar cistern. Subsequent magnetic resonance imaging and angiogram (MRI/MRA) demonstrated a 5cm, avidly enhancing lesion arising from the superior nasal cavity and extending into the sphenoid and ethmoid sinuses. The lesion involved the left

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Figure 1. MRI with avidly enhancing nasopharyngeal mass

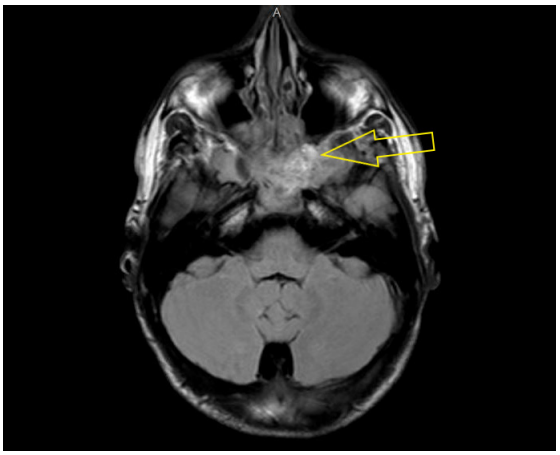


Figure 2. MRI with left sided FLAIR enhancement suggestive of central haemorrhage contained within nasopharyngeal mass

infratemporal fossa and clivus and included a 3mm vascular aneurysm. No other intracerebral vascular abnormality was identified.

Treatment

The patient was transferred to a tertiary neurosurgical centre where he completed a two week course of ceftriaxone. A tissue biopsy confirmed a diagnosis of JNA. The lesion had provided an entry point for the streptococcal pathogen from the nasopharynx, facilitated a subarachnoid haemorrhage due to its vascularity and presumably had been the cause of the patients many years of epistaxis. Treatment involved embolization and surgical resection with a good outcome. Several years on, the patient remains well and annual scans have shown no recurrence.

Discussion

Whilst JNA could be considered a paediatric pathology; the age group affected can bridge the paediatric to adult population; and as in this case patients can present to acute medical units with a condition that adult clinicians may have limited knowledge of.

Bacterial meningitis is a medical emergency with significant mortality[1] and long term neurological sequelae.[2] Although rare,[3] true bacterial meningitis can cause concern on the medical take and should prompt rapid antibiotic treatment. As in this case; in adults the most common identified causative organism is Streptococcus pneumoniae; Neisseria Meningitidis and Listeria also contribute.[4] Predisposing factors such as malnutrition, sickle cell disease and splenectomy are identified in the literature.[5] It is assumed that meningitis is preceded either by a high-grade bacteraemia, or that pathogens gain access to the CSF by dural defects or from adjacent local infections.[6] The presence of a highly vascularised abnormality between nasopharynx and subdural space provides ample opportunity for invasion, and so it is not surprising that meningitis is recognised as a potential complication of JNA.

JNA is a locally aggressive vascular neoplasm arising in the nasopharynx. First described in

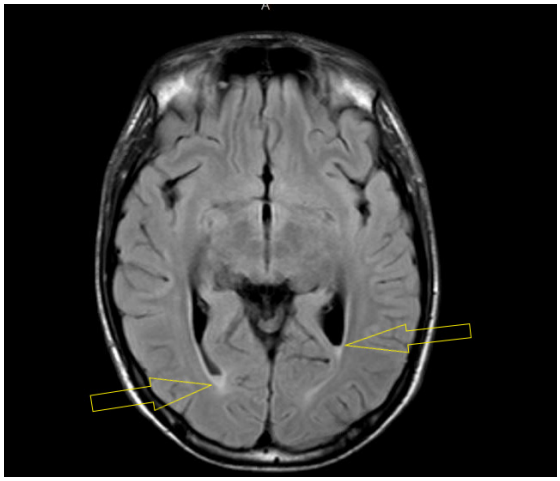


Figure 3. MRI showing subtle FLAIR-intense fluid in lateral ventricles - sub arachnoid haemorrhage.

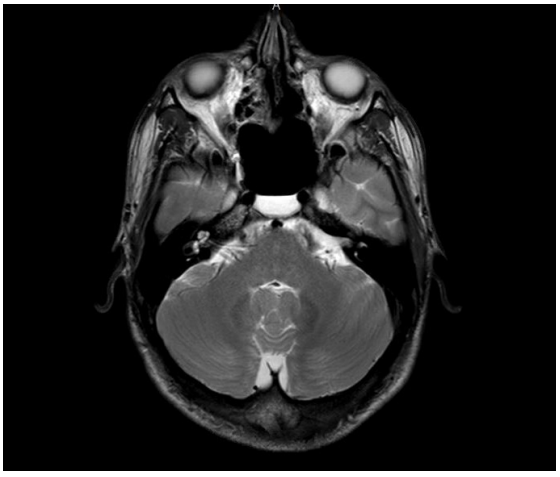


Figure 4. Figure 4 MRI 4 months post operatively showing successful JNA removal

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1940, JNA accounts for 0.05% of all head and neck cancers and almost exclusively effects adolescent males.[7] Histologically, JNA derives from vascular endothelial cells and some studies have identified the influence of vascular endothelial factor.[8] However, the role of hormone receptors remains controversial despite JNA's apparent gender and age selectivity.[7] From its point of origin, JNA tends to spread destructively into local sinus and skull base structures and so can present with a spectrum of symptoms. Typically, JNA presents as a painless, progressive unilateral nasal obstruction.[9] Headache, episodes of epistaxis, persistent rhinorrhoea and sometimes pain can also occur.

On CT scanning, JNA appears as a hetero-dense mass, enhancing with contrast.[10] MRI better identifies invasion of JNA intracranially and is useful in distinguishing JNA from other nasopharyngeal lesions in the absence of histology.[11] JNAs are treated with surgery when possible; with pre-operative embolization shown to reduce haemorrhage.[12] Recurrence is unfortunately quite common.[9]

In the context of meningitis, the role of radiological imaging remains controversial. In the absence of reduced GCS, age > 60, seizure, immunocompromise or neurological deficit evidence suggests a CT scan is not required prior to LP.[13,14] Indeed, the facilitation of a CT has been shown to delay both lumbar puncture and antibiotic administration.[14] Despite this, the trend for performing CT prior to LP is increasing,[15] likely as a result of increasing out of hours CT services.[16]

Although the patients presentation was one of infective meningitis, a xanthochromia was sent by the doctor completing the procedure probably as a matter of routine. In this instance, subsequent CT imaging revealed both blood in the subarachnoid space, but also drew attention to the JNA that may have resulted in a recurrence of meningitis in the future.

Declaration of Conflict Interest

The authors have no conflicts of interest to declare

References

1. Swartz M. Bacterial Meningitis - A View of the Past 90 Years. *New England Journal of Medicine.* 2004; **351:** 1826-1828.
2. Weisfelt *et al*. Dexamethasone and long term outcome in adults with bacterial meningitis. *Annals Neurology*. 2006; **60:** 456-68.
3. Okike IR. Trends in bacterial, mycobacterial, and fungal meningitis in England and Wales 2004-11: an observational study. *Lancet Infectious Diseases*. 2014; **14:** 301-7.
4. Durand ML, Calderwood SB, Weber DJ, *et al*. Acute Bacterial Meningitis in Adults. A review of 493 episodes. *N Engl J Med*. 1993; **328:** 21-8
5. Kastenbauer SP. Pneumococcal meningitis in adults: spectrum of complications and prognostic factors in 87 cases. *Brain*. 2003; **126:** 1015-25.
6. Hoffman OW. Pathophysiology and Treatment of Bacterial Meningitis. *Therapeutic advances in Neurological Disorders*. 2009; **2:** 1-7.
7. Coutinho-Camillo CM, B. M. Genetic alterations in juvenile nasopharyngeal angiofibromas. *Head and Neck*. 2008; **30:** 390-400.
8. Nonogaki S, C. H. Markers of vascular differentiation, proliferation and tissue remodeling in juvenile nasopharyngeal angiofibromas. *Exp Ther Med*. 2010; **1:** 921-6.
9. Makhasana JA. Juvenile nasopharyngeal angiofibroma. *Journal of oral and maxillofacial pathology*. 2016; **20:** 330.
10. Ludwig BJ, F. B.-A. Diagnostic imaging in nontraumatic pediatric head and neck emergencies. *Radiographics*. 2010; **30:** 781-99.
11. Lloyd G H. D. Juvenile angiofibroma: Modern imaging and its influence on the surgical treatment of juvenile angiofibroma. *Journal of Laryngology and Otology*. 1999; **113:** 43-4.
12. Shenouda K. Y. Endoscopic Resection of Pediatric Skull Base Tumors: An Evidence-Based Review. *Journal of Neurological Surgery: Skull Base*. 2019; **80:** 527-39.
13. Gopal A. W. Cranial computed tomography before lumbar puncture: a prospective clinical evaluation. *Archives Internal Medicine*. 1999; **159:** 2681-5.
14. Hasbun R. A. Computed Tomography of the head before LP in adults with suspected meningitis. *New England Journal of Medicine*. 2001; **345:** 1727-33.
15. Ganseshan D. CT before lumbar puncture in meningitis-what every radiology trainee should know. *Clinical Radiology*. 2009; **64:** 845-6.
16. Nagra I. W. The role of cranial CT in the investigation of meningitis. *JRSM Short Reports*. 2011; **2:** 20.

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