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Bilateral internal jugular vein thrombosis in a child with protein S deficiency

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Abstract
Internal jugular vein thrombosis is a rare but treatable disorder with most of the reports in the literature related to the adult population. The most common reported cause is iatrogenic trauma to the internal jugular vein and only a few cases have been reported in the paediatric population. We present a case of bilateral internal jugular venous thrombosis with pulmonary and cerebral complications in a 14-year-old girl with protein S deficiency. This problem has not been previously reported in the world literature.

Key words: Jugular Veins; Thrombosis; Child; Protein S

Introduction

Acute neck swellings are a common presentation particularly in children. Spontaneous internal jugular vein thrombosis, although a rare cause for an acute neck swelling, can potentially cause serious and life-threatening complications.

Case Report

A 14-year-old girl presented with a two-week history of left-sided neck pain and fever. One month previously, her general practitioner, having made a presumptive diagnosis of left-sided otitis media, had treated her with a course of amoxycillin. Prior to this event she was fit and healthy with no previous hospital admissions. A family history of protein S deficiency was noted.

On examination, she was pyrexial with a tachycardia and was slightly drowsy but fully oriented. Her neck was flexed and rotated to the left side. There was fullness over both sternocleidomastoid muscles that were tender to palpation. Otological examination was normal. Fundoscopy confirmed the presence of bilateral papilloedema and a chest examination revealed decreased air entry, stony dullness on the left side and crepitations on the right. There were no other abnormal neurological, cardiovascular or abdominal findings.

Haematological investigations showed a markedly increased white cell count (37.3 x 10^9/l) with a neutrophilia (33 x 10^9/l) and a raised c-reactive protein (CRP) (320 mg/l). Thrombophilia screening indicated a low protein S level of 31 units (normal 70–120 units). A chest radiograph showed widespread patchy consolidation and bilateral pleural effusions (Figure 1). An ultrasound examination of the neck revealed bilateral internal jugular vein thrombosis. A subsequent computed tomogram (CT) scan of the head, neck and chest confirmed thrombus in internal jugular veins extending to the sigmoid, transverse (Figure 2) and left cavernous sinuses with bilateral pleural effusions. Intravenous therapy was commenced with penicillin, flucloxacillin and heparin. Blood cultures grew *Streptococcus*, which was sensitive to penicillin.

Over subsequent days her level of consciousness deteriorated and magnetic resonance angiography (MRA) and magnetic resonance imaging (MRI) of the brain were performed. These revealed extensive venous thrombus on

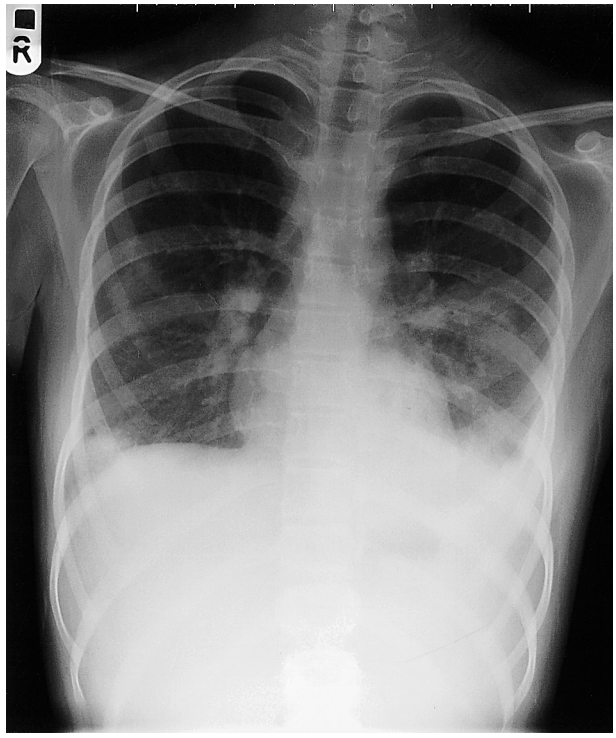

FIG. 1
Chest radiograph showing bilateral widespread patchy consolidation and pleural effusions.

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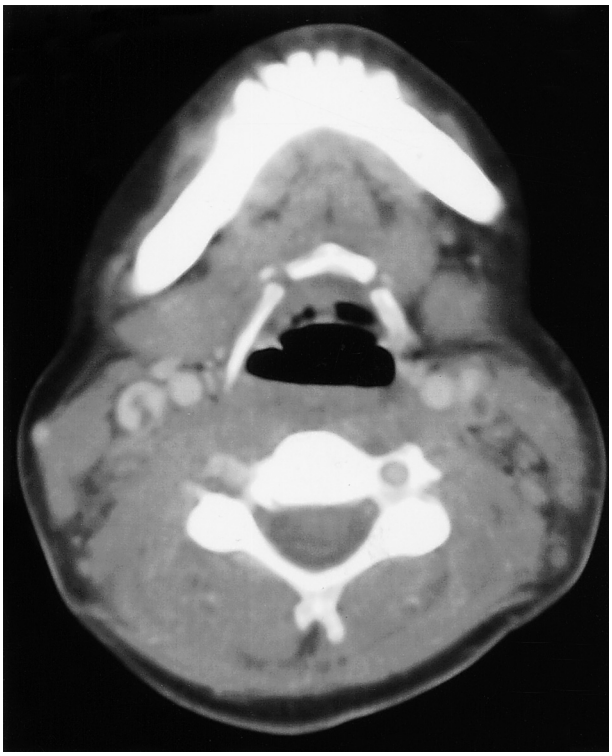

FIG. 2

Contrast computed tomogram scan of the neck showing filling defect in both internal jugular veins consistent with thrombus formation.

the left side with obliteration of the left transverse and sigmoid sinuses with near total occlusion of the right transverse and sigmoid sinuses (Figure 3). It also showed ischaemic changes in the left temporal lobe, corpus striatum and the right thalamus.

Her temperatures continued to spike and a recurrent left-sided neck swelling appeared on the 17th day. A further MRI examination of the neck and brain showed a collection deep to the trapezius muscle at the level of the

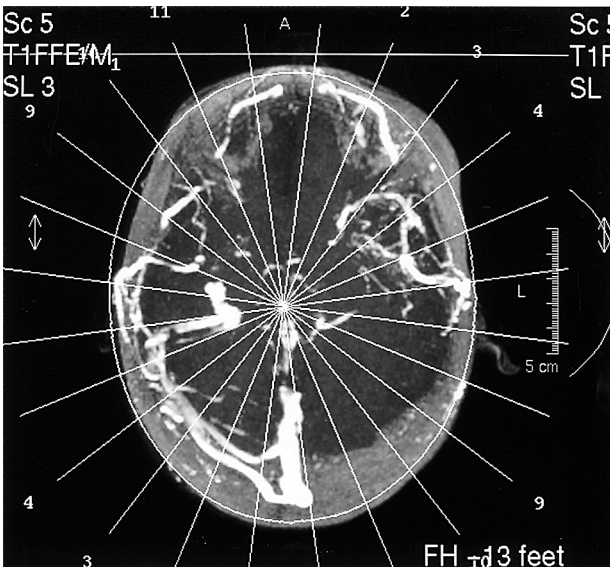

FIG. 3

Magnetic resonance angiogram showing absence of flow in the left transverse and sigmoid sinuses and diminished flow in the right anterior transverse and sigmoid sinuses.

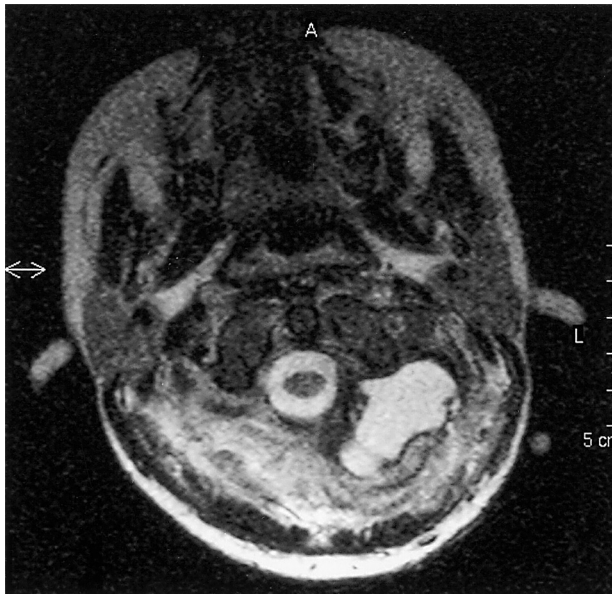

FIG. 4

T2-weighted transverse magnetic resonance imaging scan of the neck illustrating left-sided collection with surrounding oedema deep to the trapezius at the level of the craniocervical junction.

craniocervical junction (Figure 4) with surrounding oedema and multiple ring enhancing lesions in the brain (Figure 5). Haemorrhagic fluid was aspirated from the neck swelling: no organisms were isolated on culture.

In view of the spiking temperatures and recurrent neck swelling, Meropenem, a broad spectrum, beta-lactam antibiotic was added to the treatment regimen with a good

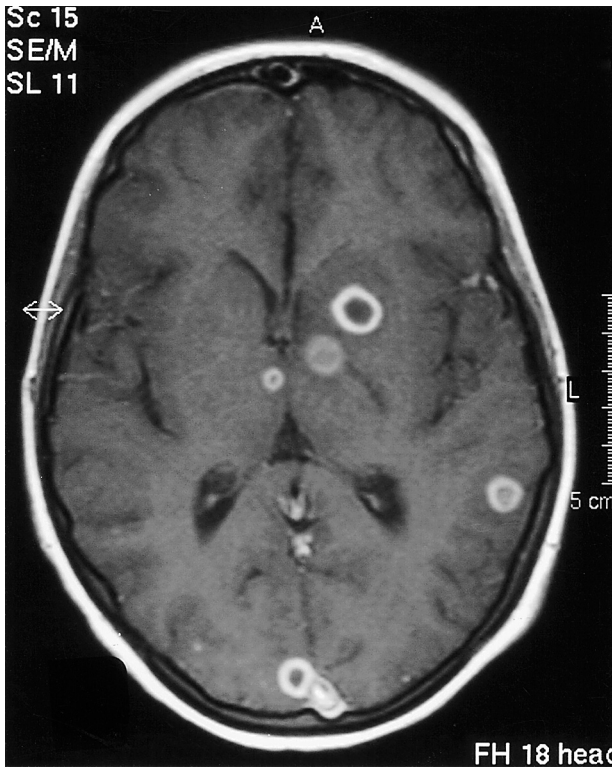

FIG. 5

Post-gadolinium T1-weighted magnetic resonance imaging of the brain illustrating multiple ring enhancing lesions.

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response and a significant improvement in her condition in the following eight-week period. Her neurological status returned to normal and the neck swelling subsided without a need for surgical intervention. Chest X-ray appearances were much improved with only a slight residual pleural thickening. A repeat MRI showed complete resolution of the thrombus with a marked regression in the cerebral ring lesions. She was allowed home with oral antibiotics and warfarin. A further eight weeks' follow up with an MRI of the brain showed complete resolution of all the cerebral lesions. Currently she is on a prophylactic dose of warfarin and regular follow up is planned.

Discussion

Internal jugular vein thrombosis is a rare, potentially fatal condition. In the pre-antibiotic era the condition was commonly related to anaerobic infections of the head and neck region. In 1936 Lemierre[1] described the condition related to the microbial organism *Fusobacterium necrophorum* which also causes oropharyngeal infections. Whilst the use of antibiotics has significantly reduced the incidence of Lemierre's syndrome, there are still some sporadic reports of this condition.[2,3]

In the present era, internal jugular vein thrombosis is more often due to direct trauma to the vein[4] by catheterization, repeated injections by intravenous drug abusers and neck dissections. Other causes are neoplasm in the head and neck region including occult disease[5] and, rarely, hereditary thrombophilic conditions complicated by ovarian hyperstimulation and *in vitro* fertilization.[6]

In the paediatric population venous thrombosis is very rare. A prospective study[7] has shown that central venous catheterization in neonates is associated with an increased risk of thrombosis in the neck, thorax, and abdomen and, in older children, additional risk factors arise if there is co-existent heart disease, immobility and surgery. Congenital prothrombotic disorders are more often found to be associated with venous thrombosis in older children compared with neonates. Unilateral internal jugular vein thrombosis has also been reported due to a pencil injury to the palate.[8]

Septic thrombi from the sigmoid sinus can propagate downwards to involve the internal jugular vein. Sigmoid sinus thrombosis is a well-known life-threatening complication of acute mastoiditis. The prevalence of sigmoid sinus thrombosis, which was mostly seen in children in the pre-antibiotic era, has fallen steeply with the introduction of antibiotics for ear infections. However, more cases are now being seen with a less overt presentation.[9]

In this patient, a history of suspected otitis media could be important in the pathogenesis of internal jugular vein thrombosis. However, given the simultaneous bilateral presentation of the thrombosis, protein S deficiency is the most likely aetiological cause in this case. Hereditary thrombophilia is due to congenital deficiencies or abnormalities of natural anticoagulant protein anti-thrombin III, protein C and protein S. The deep veins of the lower limbs are commonly involved; however, unusual sites such as the mesenteric vein and upper extremity venous thrombosis are a characteristic of hereditary thrombophilia and the risk of developing thrombosis in individuals with inherited thrombophilia is three times higher than the control group with normal coagulation.[10]

A high index of suspicion is necessary to make the diagnosis. Ultrasound or Doppler examination of the neck may reveal the thrombus in the jugular vein and CT scanning will further define the thrombus as well as revealing the extent. In recent years MRI has superseded CT scan as the investigation of choice[11] and venography is now rarely used. A full blood count, coagulation and thrombophilia screening remain essential initial investigations.

Currently, the management of internal jugular vein thrombosis is conservative with antibiotics and anticoagulants.[5] Intravenous antibiotics, particularly penicillin or third generation cephalosporins, are administered until the culture and sensitivity results are available. Controversy exists over the role of anticoagulants, in particular where the cause of internal jugular vein thrombosis is due to infection,[2,3] as it is argued that there is a theoretical risk of clot break up and distal dissemination of septic emboli. Some authors routinely recommend anticoagulants to reduce the risk of pulmonary embolism[5] while others limit its use to cases where a septic embolus persists or where there is cavernous sinus involvement.[3] Fibrinolytic agents, e.g. streptokinase and urokinase, have been successfully used[4] in adults, but are alleged to carry a greater risk of haemorrhage compared with heparin.

Surgery by ligation and excision of the affected vein was the primary treatment for internal jugular vein thrombosis before the advent of antibiotics, but ligation of the internal jugular vein is now rarely performed and is reserved for patients in whom the risk of thromboembolism persists despite adequate antibiotic and anticoagulation treatments. At present, surgery is limited to the drainage of neck abscesses.[3]

Treatment of venous thromboembolism in the paediatric age group is similar to adults. In the two reported paediatric internal jugular vein thrombosis cases, one had antibiotics without anticoagulants in addition to surgical drainage of a neck abscess[2] while the other was treated with antibiotics and anticoagulants.[8] The presence of hereditary thrombophilia with a strong family history increases the risk of recurrent thrombosis and therefore, long-term prophylactic anticoagulants should be considered.

In conclusion, bilateral internal jugular vein thrombosis is very rare in childhood. Internal jugular vein thrombosis may be the presenting feature of an underlying thrombophilic condition triggered by a mild ear or oropharyngeal infection, and this should prompt investigation into the patient's coagulation status.

The possibility of jugular vein thrombosis in children presenting with acute painful neck swellings, particularly where the initial diagnosis is not obvious, must always be considered.

- **Internal jugular vein thrombosis is most commonly unilateral and occur mostly in an adult population**
- **In the case reported here the problem arose in childhood and was bilateral and, seemingly, was the result of a relatively minor infection in association with a thrombotic condition arising as a result of protein S deficiency**
- **The management of such cases is discussed**

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