Schistosomiasis of the appendix

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Schistosomiasis of the appendix was first described in 1909, but remains a rare condition, although it has been reported in endemic areas. Increasing globalization has made such diseases important in the west.

CASE HISTORY

A 30-year-old male UK resident originally from Ghana, was admitted as an emergency with a 1 day's history of right iliac fossa pain. This had started very suddenly, initially centrally, with radiation to the right iliac fossa. There was a distinct history of anorexia but no associated nausea or vomiting. The patient denied any obvious urinary symptoms. He was otherwise, a fit and healthy person with no concurrent medical problems.

On examination he was noted to be apyrexial, tachycardic (100/min) and haemodynamically stable (148/90). Abdominal examination revealed localized peritonitis in the right iliac fossa. There was no obvious organomegally. A confident diagnosis of appendicitis was made and the patient managed accordingly. He had a negative urinalysis, a white cell count of 10.6 (75% neutrophils) and normal urea and electrolytes and liver function tests. At appendicectomy a few hours later, he was noted to have an acutely inflamed appendix with no visible perforation. He made an uneventful post operative recovery and was discharged 2 days later, pending an outpatient review 2 weeks later.

Macroscopic examination of the appendix specimen showed its surface covered by fibropurulent material but no obvious perforation. Histology showed luminal pus in the appendix associated with numerous schistosoma (*Schistosoma mansoni*), transmurally and within the subserosal adipose tissue. He was referred to a tropical disease physician to ascertain whether further treatment was required, in light of the long-term risk of portal hypertension and liver cirrhosis.

DISCUSSION

Schistosomiasis is a water-borne trematode infestation, and is one of the most widespread parasitic diseases in the world. However, it occurs in well-defined geographical areas.[1]

Schistosomiasis of the appendix was first described by Turner in 1909 (3), and has been reported in endemic areas. The most usual organisms are *Schistosoma haematobium* and *S. mansoni*.[1–4]

The actual role of infestation as a contributing factor to the development of appendicitis is still open to debate and has been the subject of much controversy. Most recently, studies in rural Egypt and West Africa, where schistosomiasis is endemic, have shown appendicitis to be relatively uncommon; and the cause is not usually attributable to a schistosomal infestation of the appendix.[3]

The characteristic pathological tissue response is believed to be a granulomatous inflammatory reaction to the schistosomal ova, with the lesion predominantly in the submucosa and serosa. There is formation of epitheloid cell granulomas which ultimately undergo fibrosis. Intramuscular oviposition (submucosa) causes an obstructive type of appendicitis with a greater risk of perforation. Serosal involvement causes inflammation and the formation of adhesions.[2,4–6]

Aside from the case report described above, two other clinical aspects of Schistosomal appendicitis are important.

First, it has been reported that schistosomiasis is a very common complication of pregnancy in endemic areas. This has been ascribed to the congestion of the pelvic vessels during pregnancy, which facilitates the passage of eggs into the intestinal villae and intervillous spaces with lodgement in the appendix and subsequent inflammation.[3]

Secondly, acute appendicitis with pseudotumerous appendix and lymph node masses has also been described at operation, even in instances where there are poor epidemiological findings on clinical history. Frozen section biopsies in these cases, may help to establish diagnosis and thus avoid unjustified bowel resection.[6,7]

Confirmation of appendicular schistosomiasis is a purely histological diagnosis, because there are no pathognomonic clinical or operative findings. It is clear that our patient required a multidisciplinary approach to his management, involving both the surgeon, and the physician.

Schistosomiasis, in endemic areas, is a major public health problem. Specific drug treatment involves the use of antihelmintics. Praziquantel is in most common usage, treating both urinary and gastrointestinal forms of the disease, and administered as a single dose.

Physicians practising in the West must be aware of the possibility of seeing atypical presentations of parasitic diseases, especially considering the worldwide increase in immigration and tourism.

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Bilateral cavernous sinus thrombosis complicating sinusitis

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Cavernous sinus thrombosis is a rare yet potentially fatal and debilitating condition in children which is important to diagnose early. It requires a high index of suspicion and confirmation by imaging.

CASE HISTORY

A previously healthy, 6-year-old boy (Figure 1) presented to accident and emergency with a 1-week history of being unwell. He complained of a 1-week headache, which radiated from the occipital to the frontal region. It was not relieved by analgesia and he remained lethargic. He had a 5-day history of pyrexia not resolving. He also had a 1-day history of redness and swelling of the left periorbital area.

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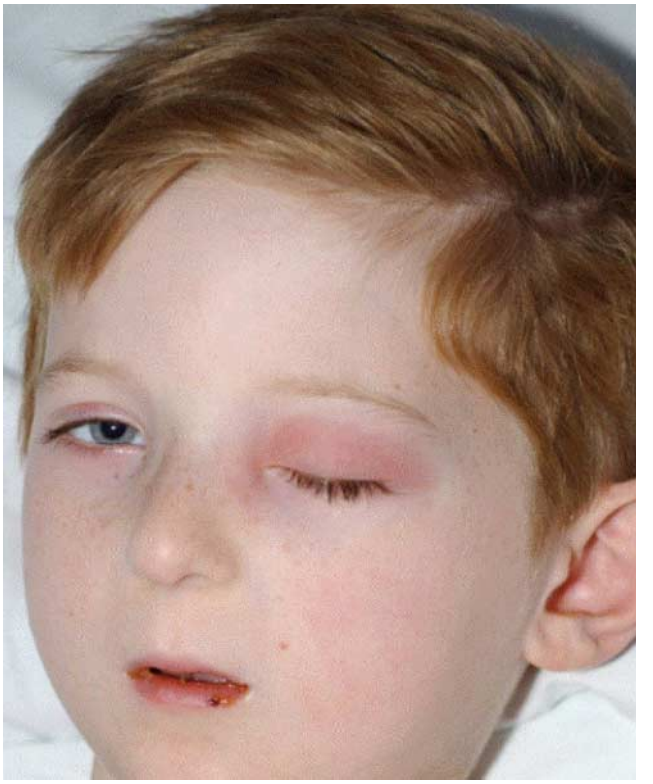

Figure 1 **Cavernous sinus thrombosis in 6-year-old boy. [In colour online]**

Examination revealed him to be irritable but with a Glasgow coma score of 15/15. He had puffiness around his left periorbital region with erythema. He had a full range of left eye movements, and pupils were equal and reactive to light. There was no involvement of his cranial nerves with normal external ocular eye movements. He otherwise had an unremarkable systemic and neurological examination.

He was commenced on intravenous antibiotics (cefuroxime and metronidazole). An initial diagnosis of preseptal cellulitis secondary to sinusitis was made. Thirty-six hours after admission it was felt that there was no improvement and he persisted to be lethargic and pyrexial so a computerized tomography (CT) scan was performed. The initial CT scan revealed bilateral cavernous sinus and left orbital vein thrombosis, sphenoid and ethmoidal sinusitis and a left prespetal cellulites (Figure 2).

He was therefore transferred to a tertiary centre for further neurological assessment and management. Anticoagulation with low molecular weight heparin was commenced. This was then changed to warfarin with the aim that his international normalized ratio should be maintained at three. His temperature continued to spike and his blood cultures on admission grew *Streptococcus milleri*. Magnetic resonance scan (MRI) of his sinuses was performed and the cavernous sinuses remained bulky. In view of this, an endoscopic sphenoidectomy and posterior

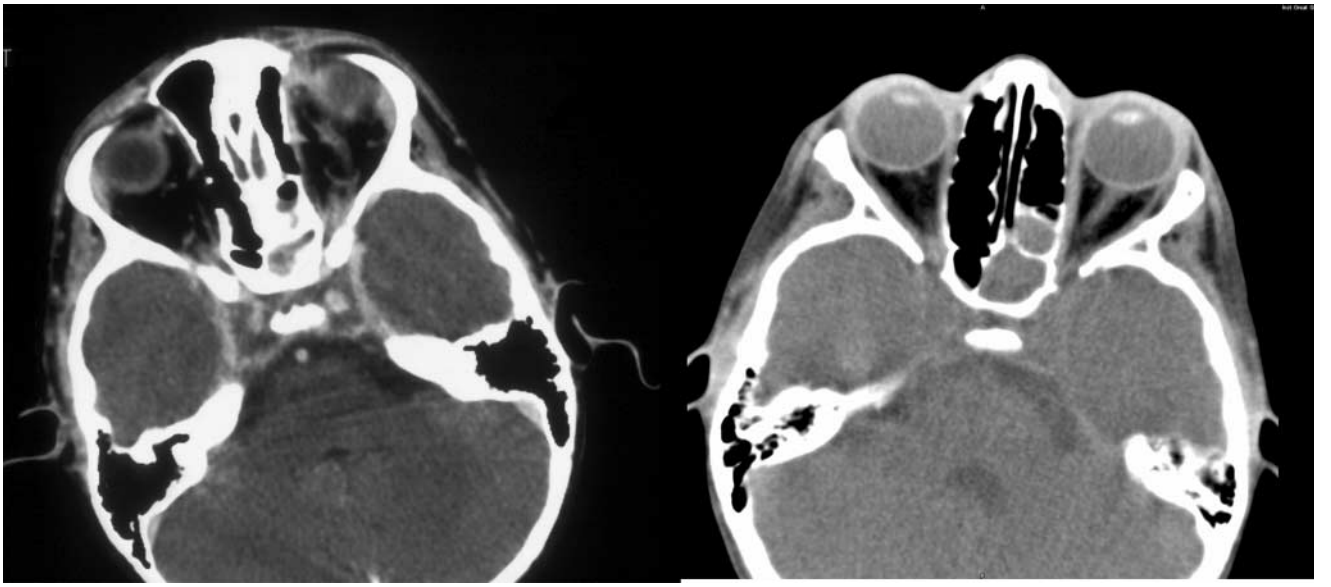

Figure 2 **The post contrast computerized tomography (CT) scan on the left shows poor enhancement of the cavernous sinus bilaterally. The uncontrasted CT scan on the right shows subtle hypodensity in the region of the cavernous sinus. There is also associated opacification of the sphenoid sinus and posteriorly situated ethmoidal air cells**

ethmoidectomy was performed and a substantial amount of pus was drained from the sphenoid ostium.

A repeat MRI three months later revealed resolution of the thrombosis and anticoagulation was stopped. He made an uneventful recovery and had no neurological sequelae.

COMMENT

The advent of antibiotics, while apparently not lessening the frequency of sinusitis, has reduced the severity and complications. Children are at greater risk of developing serious life-threatening complications from oropharyngeal infections. Early recognition and treatment are essential to reduce any subsequent morbidity or mortality. The most feared complication is intracranial suppuration, which may manifest as cavernous sinus thrombosis. In a recent review, only one child out of 104 admitted with complications of acute sinusitis had cavernous sinus thrombosis.

Cavernous sinus thrombosis may result from any infection of the tissue drained by the cavernous sinus. This includes the midface, orbit and sinonasal cavity. The mortality rate remains high at 30%, and significant morbidity includes residual cranial nerve palsies and blindness.

Thrombophlebitis of the ophthalmic veins attributable to orbital inflammation secondary to ethmoiditis may extend posteriorly, causing cavernous sinus thrombosis. The sphenoid sinus, like the frontal, develops in diploic bone and has venous communications with the adjacent cavernous sinus. Therefore sphenoiditis can result in cavernous sinus thrombosis via this route.

Clinical features vary. The onset is usually abrupt, with unilateral periorbital edema, headache, photophobia, and proptosis. In our patient, preseptal cellulitis resulted from untreated sinusitis and represented local spread of inflammatory elements to the lid. Headache and lethargy were the marked features, which were persistent after 36 h from admission. Even though there were no neurological signs, his persistent symptoms raised the index of suspicion of an intracranial complication being present and in particular cavernous sinus thrombosis. Despite its intimate connection with the cranial nerves III, IV, V, and VII, neurological dysfunction might not be typical in the early stage of the illness. This highlights the difficulties of making a diagnosis based on clinical examination alone.

The principal bacterial agents causing sinusitis are *Streptococcus pneumoniae,* other streptococci, *Staphylococcus aureus,* and non-spore-forming anaerobes. Our patient grew *S. milleri* from his cultures, which is a common pathogen with complications of sinusitis in children. Other aerobic and gram-negative organisms have also been isolated from orbital infections. Polymicrobial infections are common. The incidence of orbital and preseptal cellulitis due to *Haemophilus influenzae* has decreased dramatically over the last 10 years, owing to widespread immunization.

Most cases of paediatric paranasal sinusitis do not require radiographic confirmation and are treated medically. If an intracranial complication is suspected, imaging is required. Emergent cross-sectional imaging should include contrast-enhanced axial CT (with coronal reconstructions) to evaluate the sinuses and orbits. MRI is required in addition to this if intracranial extension is suspected. There is no role for plain radiography in the evaluation of the complications of acute sinusitis and preseptal cellulitis in a child.

Treatment for cavernous sinus thrombosis includes high-dose intravenous antibiotics directed at the most common causative organisms, coupled with surgical drainage of the primary source of infection.

Our case highlights that the diagnosis of cavernous sinus thrombosis complicating sinusitis and periorbital cellulitis requires a high index of suspicion and confirmation by imaging. Examination may normal early on in this rare disease, and early multi-disciplinary team (ENT, ophthalmology, and radiology) working is vital. Preseptal cellulitis is a common presentation in children, and the possibility of underlying sinusitis and its complications should be thought of.

Competing interests None declared.

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