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Case Report

Lemierre syndrome: Remember the forgotten disease

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Lemierre syndrome is a rare clinical entity which is characterised by septicaemia arising from a head and neck source, complicated by thrombosis of the ipsilateral internal jugular vein with embolisation to various sites, most usually the lungs. It is important to diagnose the syndrome correctly since its treatment is different from pulmonary emboli and uncomplicated ENT infections and if untreated may be associated with high mortality. We present the occurrence of this syndrome in a 14 year-old girl whose site of primary infection was the middle ear; the outcome in this case was favourable.

INTRODUCTION

Lemierre syndrome, described as the 'forgotten disease'[1] although very uncommon[2] is an infection-related syndrome which is important to diagnose accurately since its mortality may be high.

CASE REPORT A 14-year old girl, with a history of chronic mastoiditis, who gave a three-week history of left otalgia and headache presented to hospital with worsening pain, dizziness, vomiting and rigors following seven days of oral amoxicillin for presumed acute otitis media. She was febrile and had cervical lymphadenopathy with neck pain and tenderness. A diagnosis of acute suppurative otitis media was made for which she received intravenous cefotaxime 2g 8-hourly and metronidazole 500mg 8-hourly.

Over the following five days she had persisting fever and worsening left-sided neck pain. Furthermore, she became dyspnoeic, developing pleuritic chest pain and haemoptysis; measured oxygen saturation on room air was 88%.

Computerised tomography (CT) revealed thrombus, bubbles of gas in the left internal jugular vein (*Fig. 1*) and several ill defined

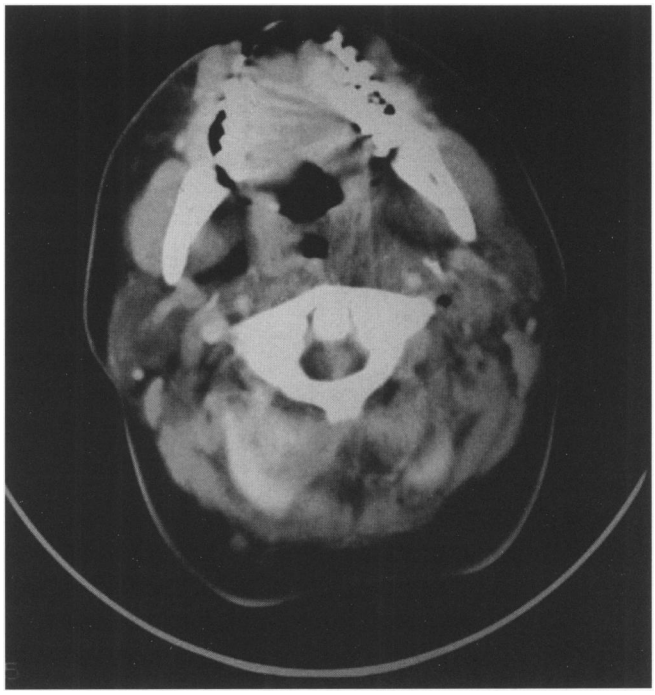

Fig 1. CT scan of neck highlighting the thrombus (manifest by the lack of contrast agent seen) in the left internal jugular vein.

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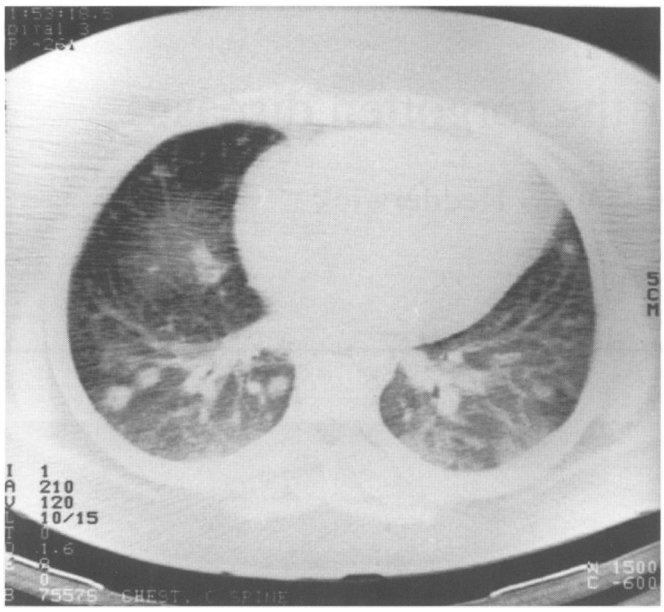

Fig 2. CT scan of chest showing several opacities with surrounding infiltrate attributed to septic emboli.

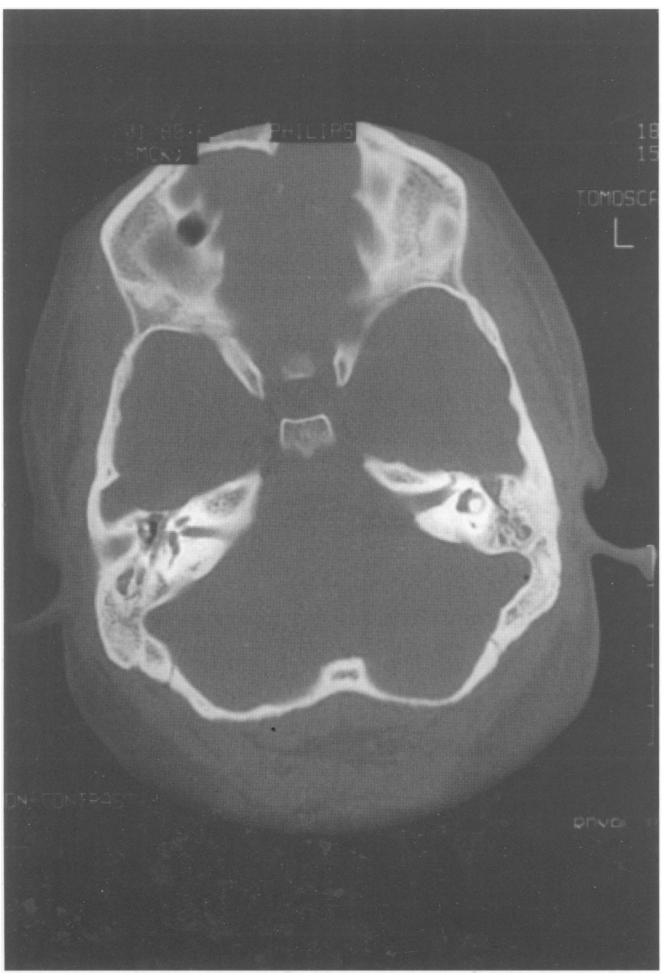

Fig 3. CT scan of head showing mastoid sclerosis, especially on left.

opacities in the lung fields consistent with septic pulmonary emboli (*Fig. 2*). Further imaging identified bilateral sclerosis of the mastoids, greater on the left, consistent with chronic mastoiditis; small gas collections and soft tissue changes consistent with acute inflammation were also present *(Fig. 3)*. Blood cultures were positive for *Bacteroides fragilis* and an unidentified anaerobic Streptococcus sp.

The patient underwent a left sided modified radical mastoidectomy and removal of granulations. She received 14 days of intravenous piperacillinl/ tazobactam 4.5g 8-hourly and metronidazole 500mg 6-hourly followed by a further 14 days of oral co-amoxiclav and metronidazole. Initial anticoagulation was with subcutaneous enoxaparin 100mg daily and, subsequently, warfarin for a total of two months. She remains well six months later.

DISCUSSION

Although the correlation between oropharyngeal infection and sepsis was first described by Courmant and Cade in 1900,[3] Lemierre was the first to characterise the syndrome in 1936 and published a series of 20 cases, 18 of whom died.[4] Alston subsequently reported a series of 280 episodes; these typically occurred in young, otherwise healthy patients.[5] The most frequent site of primary infection was the oropharynx and ranged from mild to fulminant in severity. The syndrome characteristically associates such infection with ipsilateral internal jugular vein thrombosis complicated by septic emboli, usually to the lungs, with persistent fever; the usual organism isolated from blood is *Fusobacterium necrophorum.*

Lemierre, in his original description, acknowledged alternative extrapharyngeal sources of infection which included the middle ear, female urogenital tract and gastrointestinal tract. Subsequently many sites of septic emboli have been identified such as bone, meninges, abdominal viscera, peritoneum and soft tissue.[5] Furthermore several alternative pathogens have been proposed, including *Bacteroides*, as organisms capable of precipitating the syndrome.

It is important to make the correct diagnosis to allow selection of appropriate treatment; this is different from that which is usual for pharyrigitis, otitis media, or thromboembolic disease. Debridement of the primary focus is an important

intervention and broad spectrum antibiotic therapy with potent anaerobic activity is the cornerstone of management; examples may include either clindamycin or a combination of a beta-lactam and metronidazole. The preferred duration of antibiotic therapy is 2-6 weeks, with conversion from intravenous to oral administration when marked improvement with defervescence, resolution of leucocytosis and falling inflammatory indices are observed. Mortality today is less than 17%,[6] much lower than in the preantibiotic era.

Furthermore, the role of anticoagulation remains unclear since no randomised controlled trials exist to test the hypothesis that this is beneficial. Interestingly, case series suggest that those who are anticoagulated have similar outcome to those who are not; indeed a small number suffer adverse events attributable to warfarin therapy.[7] Some have recommended that, except when cavernous sinus thrombosis is present, anticoagulation is withheld.[6]

Lemierre remarked, in his original description, that upon observation of the constellation of findings which characterise the syndrome "mistake is almost impossible".[4] However, unfamiliarity with this disease may confound the diagnosis and lead to missed therapeutic opportunity. The clinical lesson is, therefore, straightforward: when presented with a young, previously well patient who has a primary head and neck infection but a clinical illness out of proportion with this, often with symptoms at a discrete site, remember the forgotten disease.

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