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Case report

Lemierre syndrome associated with 12th cranial nerve palsy—A case report and review

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ABSTRACT

Since the widespread availability and use of antibiotics the prevalence of Lemierre syndrome (*L.S.*) has decreased. It is a well-described entity, consisting of postanginal septicaemia with thrombophlebitis of the internal jugular vein with metastatic infection, most commonly in the lungs. The most common causative agent is a gram-negative, non-spore-forming obligate anaerobic bacterium, *Fusobacterium necrophorum* (*F.n.*). We describe the unusual clinical features of a 12-year-old boy with Lemierre syndrome with isolated hypoglossal nerve palsy – the latter symptom is an extremely rare manifestation of this disease.

1. Introduction

Lemierre syndrome (*L.S.*) is a rare, but well-characterized disease, consisting of postanginal sepsis, internal jugular vein thrombophlebitis and septic emboli secondary to the infection [1]. It is mainly caused by *Fusobacterium necrophorum* (*F.n.*), a non-spore-forming, strictly anaerobic Gram-negative bacterium and affects healthy young adults. Other microorganisms implicated belong to *Fusobacterium* species, anaerobic streptococci and Gram-negative anaerobes [3]. After crossing mucosal barrier postanginal sepsis develops by haematogenous spread via tonsillar veins or directly through cervical tissue. The resulting septic thrombophlebitis of the internal jugular vein subsequently leads to metastatic lesions in different organ systems.

Although the first description of human postanginal septic infection with *F.n.* is attributed to Courmont and Cade [2] it was the French microbiologist André Lemierre who defined the clinical entity of this syndrome. From the time of *L.S.*'s initial description until the beginning of the antibiotic era in the 1940s, more than 250 cases of *F.n. bacteraemia* have been reported worldwide. From 1950 to 1995 only 40 cases of *L.S.* were presented [10]. In the 1980s, an increasing number of cases emerged; studies from 1970 to 2007 showed an incidence of 0.8–1.5 per million per year [5,6,14].

We discuss a case of Lemierre syndrome with isolated left hypoglossal nerve palsy and review therapeutic strategies for the management of this syndrome.

2. Case report

A 12-year-old boy presented with a four day-history of sore throat, neck pain and fever. A viral upper respiratory tract infection was suspected. Due to the increasing neck pain he received an occipital intramuscular injection of corticosteroid. Subsequently, neck pain increased and high fever persisted. On admission the patient had swollen cervical lymph nodes and a fixed cervical spine rotation to the left (torticollis). The oropharyngeal inspection was unremarkable without signs of retropharyngeal abscess. Antibiotic therapy with cefuroxime (100 mg/kg/d i.v.) was started as empirical

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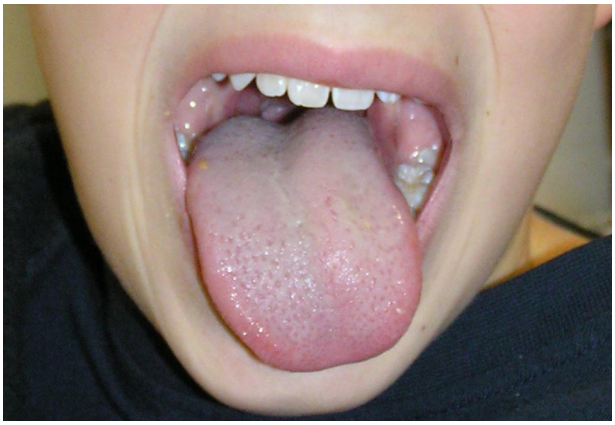

Fig. 1. Left-sided hypo-glossal palsy with deviation of the tongue at admission (day 1).

antibiotic treatment for oropharyngeal bacterial infection. On the following day he developed a left hypoglossal palsy with deviation of the tongue at clinical inspection (Fig. 1) – and clinical signs of sepsis consisting of hyperpyrexia, arterial hypotension and decreased oxygen saturation. Therefore he was admitted to intensive care unit. The laboratory results revealed thrombocytopenia (100,000/μl), coagulopathy (quick 50%), impaired kidney (creatinine 1.9 mg/dl) and liver function (AST 170 U/l, ALT 125 U/l) as well as a highly elevated C-reactive protein (206 mg/l). White blood cell count and lymphocytic differentiation were normal. A MRI scan showed inflammatory lesions dorsally and laterally to the left occipital condyle in addition to suspected thrombophlebitis of the left jugular internal vein (Figs. 2 and 3). The chest X-ray showed bilateral pulmonary lesions (Fig. 4). The patients needed supplemental oxygen and dopamine therapy. After *L.S.* was suspected the antibiotic therapy was changed to high dose meropenem (200 mg/kg/d i.v.) and clindamycin (40 mg/kg/d i.v.). Two days later, after transient improvement of his clinical condition, fever

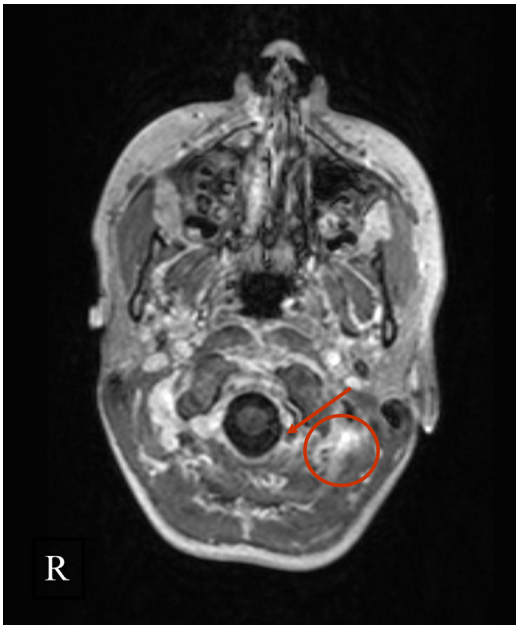

Fig. 2. Magnetic resonance imaging of the brain on day 1, transversal, T1, with contrast. Red circle: inflammatory lesions near the left occipital condyle and the hypoglossal canal. Red arrow: region of the hypoglossal canal. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

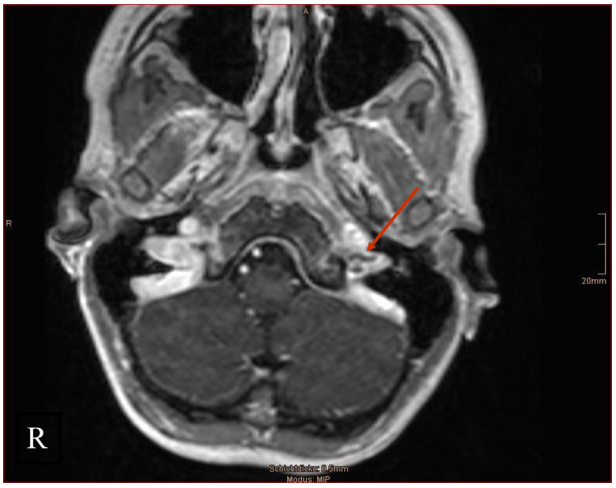

Fig. 3. Magnetic resonance imaging of the brain on day 1, transversal, T1, with contrast. Red arrow: filling defects of the left internal jugular vein as a sign of thrombosis. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

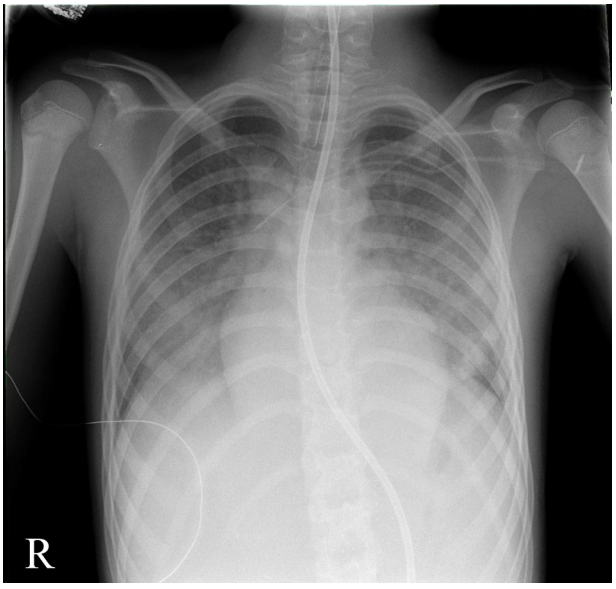

Fig. 4. Chest X-ray on day 3: bilateral pulmonary infiltrates.

recurred with increasing neck pain, a left-sided cervical swelling, and severe earache. The patient's general condition deteriorated. Due to worsening hypoxemia, the patient required intubation and mechanical ventilation.

The clinical inspection showed cervical lymphadenopathy, signs of mastoiditis (local pain, redness) and otitis media detected by otoscopy, but no retropharyngeal swelling so far. CT scan revealed cervical lymphadenopathy, torticollis of the atlanto-axial joint (Fig. 5) and fluid collection within the left mastoid air cells and the middle ear (Fig. 6). A retropharyngeal hypodensity was suspected by radiologist. During mastoidectomy and paracentesis intraoperative inspection revealed no findings of retropharyngeal abscess. Therefore no additional neck surgery was performed. After surgery the patient showed a significant clinical recovery with ongoing antibiotic treatment. Surgical cultures as well as initial blood cultures remained sterile. *F.n.* was detected by PCR from the surgical cultures. Low-dose heparin therapy was started. Within