Lemierre Syndrome in the Antibiotic Era

Lemierre syndrome is caused by an acute oropharyngeal infection associated with secondary septic thrombophlebitis of the internal jugular vein, frequently complicated by metastatic infections to the lungs or large joints.

A previously healthy and fully immunized 1½ year old girl was admitted with fever, neck pain and moderate dyspnea for four days. On examination, there was bilateral anterior cervical lymphadenopathy and tender swelling in the left laterocervical region extending from the angle of jaw and parallel to the sternocleidomastoid muscle. Laboratory analysis showed a non specific inflammatory reaction with leucocytosis and elevated ESR. Throat cultures were negative for *Corneybacterium diphtheriae* and beta hemolytic streptococci. The chest *X*-ray showed bilateral perihilar infiltrates indicating metastatic infection from oropharynx. Color doppler ultrasonography of the neck revealed thrombophlebitis of left internal jugular vein. A diagnosis of Lemierre syndrome was made. The child was treated with crystalline penicillin and chloramphenicol, metronidazole and low molecular weight heparin. Blood cultures were sterile. Serial radiologic follow up revealed resolution of the thrombus over time.

Fusobacterium necrophorum is the etiological agent in over 80% of cases of Lemierre's syndrome(1). We did not isolate this organism, as we had already started antibiotics before the blood culture was taken. The palatine tonsils and peritonsillar tissue are the primary source of infection in the majority, although pharyngitis, otitismedia and mastoiditis have been described(2,3). Lungs are the most common sites of embolic disease(3). A tender swelling at the angle of the jaw and parallel with the sternocleidomastoid muscle reflects the development of thrombophlebitis of the internal jugular vein. The mainstay of treatment is prolonged intravenous antibiotics directed at anaerobic microbes and therapeutic anticoagulation.

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Liver Transplant for Crigler-Najjar Syndrome

Crigler-Najjar syndrome is a rare genetic disorder characterized by severe indirect hyperbilirubinemia from birth(1). In Crigler-Najjar type I, where there is complete functional loss of an enzyme which glucuronidates bilirubin(2), patients usually succumb to the neurotoxicity of bilirubin early in life and if they survive, uncontrolled bilirubin levels can have detrimental effects on neurodevelopment. Liver transplantation offers the only definitive treatment. We report the first successful liver transplant for Crigler-Najjar syndrome in India.

A 27 month old girl child with indirect hyperbilirubinemia with bilirubin levels > 20 mg/dL was diagnosed as a case of Crigler-Najjar syndrome type I in a Middle East country and was discharged at 2 months of age on home phototherapy and later

started on phenobarbitone. In view of her persistent hyperbilirubinemia despite conservative therapy and severe limitation in mobility because of the need for prolonged phototherapy, she was referred to our centre for liver transplantation.

On admission she weighed 14.8 kg and was deeply icteric. The investigations revealed an indirect hyperbilirubinemia (total bilirubin 29.31 mg/dL, direct 0.53 mg/dl), AST 41 U/l, ALT 46 U/l, alkaline phosphatase 218, prothrombin time 12.4 sec. The infant received a segment II and III graft from the left lobe of her father's liver. Post operatively she remained hemodynamically stable and was extubated the next day. She recovered well and was discharged 21 days after liver transplantation. Her pre discharge investigations revealed a bilirubin of 1.8mg/dl, direct 1 mg/dl, AST 46 U/l, ALT 31 U/l, serum alkaline phosphatase 131. At fifteen months post-transplant the child is doing well with normal liver function and a serum bilirubin of 0.8 mg/dL.

This case highlights the growing acceptance of liver transplant in India for a variety of indications including metabolic disorders.

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