J Neurol (2008) 255:1588–1589
DOI 10.1007/s00415-008-0965-9

Ewoud J. van Dijk
John C. van Swieten
Peter J. Koudstaal

Meningitis, cranial nerve palsies and bilateral cerebral infarcts

A neurological variant of Lemierre's syndrome

Received: 26 December 2007
Received in revised form: 24 March 2008
Accepted: 3 April 2008
Published online: 2 August 2008

Sirs: In 1936, Lemierre reported on orophayngeal infections extending to the lateral pharyngeal space, causing septic jugular thrombophlebitis with embolisation mostly to the lungs [4]. The main aetiological pathogen is Fusobacterium necrophorum, an anaerobic gram-negative rod that is part of the oropharyngeal flora. Widespread use of antibiotics for common pharyngitis made the syndrome rare and hence even forgotten [5].

We present a previously healthy 17 year-old man with a sore throat, headache and low-grade fever without general sickness. Initially, he was treated symptomatically under the suspicion of viral pharyngitis. After six days he was referred to a local hospital with a 39 °C temperature, somnolence, dysphagia, dysarthria, painful neck and palpable cervical lymph nodes. Neurological examination showed left abducens nerve palsy, dysarthria and meningism. Blood tests revealed platelet count 3.0×10^9/L, WBC 10.2×10^9/L (95 % neutrophils) and CRP 348 mg/L. CT scan of the head showed ethmoid and sphenoid sinusitis, but no cerebral abnormalities. Lumbar puncture under platelet cover, yielded purulent CSF, with 630×10^6/L white cells (94 % neutrophils), total protein of 1.54 g/L, and glucose of 2.9 mmol/L. He was treated with dexamethasone and cephotaxim under the suspicion of bacterial meningitis. Fusobacterium necrophorum was cultured from throat smear, blood and CSF. Antibiotics were changed to ciprofloxacin and metronidazole. Seven days after admission he developed left-sided hemiparesis, while platelet count had normalised. MRI scans showed bilateral white matter lesions in the semioval center, left internal carotid artery occlusion, loss of sphenoid sinus pneumatisation, contrast enhancement of the cavernous sinus and debris in the cavum Meckeli (Fig. 1 a). After transfer to our hospital, neurological examination showed diminished visual acuity of the left eye secondary to cornea erosion, left-sided Horner syndrome, bilateral hypoesthesia in the upper two trigeminal nerve branches, bilateral abducens nerve paresis, dysarthria, left-sided paresis of the pharyngeal, vagus, accessory and hypoglossal nerves and paresis of the left leg with Babinski sign. CT angiography showed left carotid artery occlusion from bifurcation to siphon and subtotal right carotid artery stenosis at the siphon and petrosal segment, but no venous abnormalities (Fig. 1 b). Ciprofloxacin was switched to penicillin, bilateral antral wash-outs and opening of ethmoid and sphenoid sinuses were performed, aspirin was started, and the cornea erosion was treated. Virological assay showed a past Epstein-Barr virus infection. Within 3 months our patient completely recovered except for dysarthria and bilateral facial hypoesthesia. CT angiography after 9 months showed an occluded left and a severe stenotic right carotid artery (Fig. 1 c).

Pharyngitis can extend to paranasal sinuses and lateral and retropharyngeal spaces. In our patient parapharyngeal involvement resulted in palsies of sympathic and caudal brainstem nerves, clinically known as Villaret's syndrome. Cavernous sinus and cavum Meckeli involvement resulted in carotid stenosis, meningitis and trigeminal and abducens nerves palsies.

Thromboembolic complications

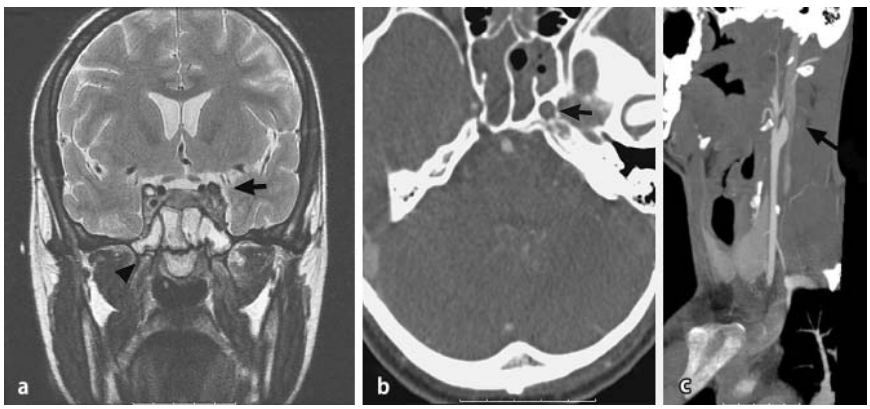

Fig. 1 **a** T2W MRI: occlusion of cavernous part of left ICA (arrow), loss of liquor content of left cavum Meckeli due to debris, loss of pneumatisation of sphenoid en ethmoid sinus (arrowhead). **b** CT angiography: left sided ICA occlusion (arrow) and right sided ICA stenosis. Loss of pneumatisation of sphenoid sinus.**c** CT angiography after 9 months: ICA occlusion just distal from the bifurcation

E. J. van Dijk, MD, PhD (✉) ·
J. C. van Swieten, MD, PhD ·
P. J. Koudstaal, MD, PhD
Dept. of Neurology
Erasmus MC
's Gravendijkwal 230
3015 CE, Rotterdam, The Netherlands
Tel.: +31-10/7033-307
Fax: +31-10/7033-208
E-Mail: e.vandijk@erasmusmc.nl

JON 2965

days after an uneventful pharyngitis or sinusitis is characteristic for Lemierre's syndrome. Instead of the known venous complications our patient developed thrombotic stenosis of both internal carotid arteries. Compression, inflammation and erosion of the carotid artery may induce endothelial dysfunction, vasospasm, platelet adhesion, dissection, and luminal occlusion [3]. Haemaglutinin produced by Fusobacterium species enhances platelet aggregation [2]. The temporal relationship between platelet count and clinical signs of cerebral infarction is striking. Possibly, recovery from consumptive thrombocytopenia preceded further cerebral thrombosis.

Platelet aggregation inhibitors are essential in the treatment of arterial thrombosis. A combination of penicillin and metronidazole for at least six weeks is probably the optimal antibiotic treatment. Surgical drainage can be of additional value. Lemierre's syndrome can be easily diagnosed on the basis of classical clinical features [5]. Antecedent infection with EBV is often mentioned as provocating bacterial infection [1]. In atypical cases with only neurological complications prompt recognition is essential to timely start adequate treatment.

Conflict of interest The authors declare no conflict of interest.

References

1. Dagan R, Powell KR (1987) Postanginal sepsis following infectious mononucleosis. Arch Intern Med 147:1581–1583
2. Horose M, Kiyoyama H, Ogawa H, Shinjo T (1992) Aggregation of bovine platelets by Fusobacterium necrophorum. Vet Microbiol 32:343–350
3. Jones TH, Bergvall V, Bradshaw JP (1990) Carotid artery stenoses and thrombosis secondary to cavernous sinus thromboses in Fusobacterium necrophorum meningitis. Postgrad Med J 66:747–750
4. Lemierre A (1936) On certain septicaemias due to anaerobic organisms. Lancet i:701–703
5. Screaton NJ, Ravenel JG, Lehner PJ, Heitzman ER, Flower CD (1999) Lemierre syndrome: forgotten but not extinct – report of four cases. Radiology 213:369–374