The Changing Face of Lemierre's Syndrome

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Lemierre's syndrome is considered to be a rare disorder, characterized by thrombophlebitis of the internal jugular vein following primary infection in the oropharynx, septicemia demonstrated by at least one positive blood culture, and presence of metastatic infections [1,2]. Although Sinave et al. [2] defined the syndrome as including the above four criteria, other cases reported in the English literature do not fulfil these four criteria, but are still characterized by internal jugular vein thrombosis following an oropharyngeal infection.

Lemierre's syndrome is traditionally characterized by a fulminant course, which is not surprising considering the presence of endovascular infection with metastatic foci. Nonetheless, the description of the syndrome, also called post-angina septicemia, dates back to the pre-antimicrobial era when prompt antibiotic treatment was simply not an option. In the current post-antimicrobial era, Lemierre's syndrome has not disappeared but some of its classical characteristics have changed.

This issue of *IMAJ* includes a case report of Lemierre's syndrome diagnosed by imaging rather than by physical examination [3]. The young patient displayed clinical deterioration following bacterial pharyngitis. The clinical picture was associated with dyspnea, chest pain and bloody mucous. The differential diagnosis in this case included infectious mononucleosis, secondary pneumonia and bacterial endocarditis. The presumably rare syndrome named after Lemierre was not considered at this stage, not only because it is a rare diagnosis but also because this patient presented an atypical form of the syndrome. Even though Lemierre's syndrome is characterized by a fulminant or severe course, as in this case, the presented patient did not have the characterizing "cord sign," an induration of the internal jugular vein beneath the anterior border of the sternocleidomastoid muscle [2]. In view of the negative blood cultures combined with the lack of evidence of internal jugular vein thrombosis in the physical examination, the clinical diagnosis of Lemierre's syndrome seemed unlikely. Nonetheless, the demonstration by computerized tomography of thrombophlebitis in a branch of the left internal jugular vein resolved the diagnostic question.

The above-described atypical presentation of Lemierre's syndrome diagnosed by imaging studies could be one of other variants of the syndrome in the post-antimicrobial era. We have described two cases of the syndrome that fulfilled two or three of the criteria for Lemierre's syndrome stipulated by Sinave et al. [2]. Neither of our patients had positive blood cultures, but they did have an oropharyngeal infection followed by swelling of the right side of their necks, which was demonstrated to be due to internal jugular vein thrombosis [4]. In one of these patients a pulmonary abscess was also found. These cases differ from those described by Ben-David et al. [3] in two important aspects. First, their patient had severe illness as compared to our two described patients who had a relatively mild clinical picture [4]. Moreover, whereas in the former case the diagnosis of Lemierre's syndrome was considered only after thrombophlebitis could be demonstrated by imaging studies, our two patients had diffuse swelling of the right side of the neck, in one of them extending from the angle of the mandible to the medial end of the clavicle. Hence, in these cases, Lemierre's syndrome was suspected based on the physical examination and only confirmed (rather than diagnosed) by ultrasound scans and computerized tomography. The relatively indolent course of these patients was attributed to the early administration of antibiotic therapy, as regular therapy of suspected bacterial oropharyngeal infection. Therefore, in the post-antimicrobial era one should expect different variants or, alternatively, incomplete forms of Lemierre's syndrome.

Another interesting, yet unanswered question regarding Lemierre's syndrome is its etiology and pathogenesis. A suggested mechanism of thrombosis is propagation of thrombophlebitis of the tonsillar veins, either via lymphatic spread or by direct involvement of the alveolar tissue of the neck, into the internal jugular vein [5,6]. However, the mystery is far from being unravelled. There is no good answer to the question why simple (in most cases) oropharyngeal infection, which resolves with or without antibiotic therapy and without sequelae in the vast majority of patients, leads to such thrombosis and a possibly life-threatening condition in the minority of patients who are otherwise healthy young subjects. One possibility is a minor immunodeficiency state, which was not searched for or recognized by the respective physicians. Alternatively, patients who develop Lemierre's syndrome could have some kind of thrombophilia, as the principal feature of this syndrome is thrombosis. Propagation of thrombophlebitis of the tonsillar veins into the internal jugular veins can cause thrombosis upon presence

of a prothrombotic state. These were unknown in past decades, and hence not searched for. The options are numerous, ranging from inherited thrombophilias affecting mainly veins, to acquired prothrombotic states such as the antiphospholipid syndrome.

In conclusion, Lemierre's syndrome – a rare disorder classically characterized by internal jugular vein thrombosis, oropharyngeal infection, septicemia and presence of metastatic infections – may be encountered today (the post-antimicrobial era) in other variants. Physicians should be aware of this syndrome and its changing face and should consider it in the differential diagnosis. Incomplete forms of Lemierre's syndrome are more likely to appear because of the wide use of antibiotics, and hence severe infections should prompt the search for occult thrombosis and vice versa. Moreover, signs of internal jugular vein thrombosis in the physical examination should also suggest Lemierre's syndrome even when the clinical course is indolent. Another challenge for physicians and researchers is identifying the underlying predisposing factors that make a patient prone to internal jugular vein thrombosis following an oropharyngeal infection.

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