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Lemierre Syndrome: A Case Report[1]

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Lemierre syndrome is a rare disease characterized by internal jugular vein thrombosis and septic emboli, and it primarily occurs in healthy young individuals; this disease usually follows an acute oropharyngeal infection. To the best of our knowledge, only a few reports about this disease have appeared in the radiologic literature. We report here the radiologic findings of a case of Lemierre syndrome in a young healthy female adolescent who had a history of acute pharyngotonsilitis. Chest radiographs showed lung nodules that displayed cavitary changes with rapid progression on the serial studies. High-resolution CT scan showed multi-focal patchy consolidations that connect with vessels, and this was suggestive of septic pulmonary embolism. Ultrasonography and CT scan of the neck revealed right internal jugular vein thrombosis.

Index words : Thorax, CT
Lung, infection
Veins, thrombosis

Lemierre syndrome is an uncommon, but potentially life-threatening disease that occurs after an episode of pharyngotonsilitis. It consists of septic thrombosis of the internal jugular vein and bacteremia, and this can lead to septic pulmonary emboli and metastatic abscesses. In the preantibiotic era, Lemierre syndrome was common and it often followed a fulminant course with a mortality rate of 90% (1). The incidence of Lemierre syndrome has significantly decreased and the outcomes have improved with the wide spread use of antibiotics (2). The prognosis of Lemierre syndrome depends on the early recognition of this syndrome and adequate treatment.

We report here on a case of Lemierre syndrome in young healthy female adolescent who developed septic pulmonary embolism.

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Case Report

A 16-year-old female adolescent was admitted to the emergency department with 3 to 4 day history of sore throat, fever, myalgia, dyspnea and pleuritic chest pain that had been treated with the oral administration of antibiotics at the local hospital. However, there was no significant improvement of her symptoms. On the physical examination, right tonsilar enlargement was observed. The results of serologic examination were leucopenia (3.6×10/L), thrombocytopenia (49×10/L), and a prolonged prothrombin time (16.9 sec), and these findings were suggestive of systemic inflammatory response syndrome.

The chest radiographs obtained at admission showed mild patchy opacity in the right middle lung zone and bronchovascular bundle thickening with mottled infiltration in both lower lung zones. Nafcillin and ceftriax-

one were initially administered intravenously. Two dimension echocardiography obtained on the day of admission revealed no significant abnormality. High-resolution CT scan showed multi-focal patchy consolidations with surrounding ground-glass attenuations in both lungs, and this was mainly seen in both lower lobes along with a peripheral predominancy. These multi-focal patchy consolidations showed connection with vessels, so their appearance was suggestive of septic pulmonary emboli. Associated interlobular septal thickening and a small amount of bilateral pleural effusion were also demonstrated (Fig. 1A). Abdomen CT was performed to figure out the cause of her fever. On this study, these multi-focal patchy consolidations showed an increase of their extent and there were varying degrees of cavitations on the lung window setting (Fig. 1B). Hepatosplenomegaly was noted in the abdomen, but the rest of the abdomen was normal. Serial chest radiographs showed increasing bilateral pleural effusion and obvious cavitary changes of the lung nodules (Fig. 1C). These radiologic findings showed rapid progression on the serial studies. The patient was transferred to the intensive care unit because her fever, dyspnea and chest pain were aggravated. The pleural effusions were drained; however any organism was not isolated. Ultrasonography of the neck was performed on the 8th day of hospitalization and it revealed a non-compressible echogenic occlusive thrombus without a color flow signal in the right internal jugular vein with venous wall thickening and segmental fusiform dilatation (Fig. 1D). The antibiotics regimen was modified so that cefazoline, aztreonam and metronidazole were now administered intravenously. A throat swab and several sets of blood and sputum cultures were taken, but any isolated organism was not obtained. Cultures for anaerobic bacteria were taken two times, but unfortunately, they didn't reveal any bacterial growth. On the 16th and 20th day of hospitalization, ultrasonography and CT scan (Fig. 1E) of the neck were performed, which showed persistent thrombosis of the right internal jugular vein, although there was improvement of the pharyngotonsilitic symptoms. The findings of septic pulmonary emboli slowly improved on the serial chest radiographs from eight days after admission and the bilateral pleural effusion was also improved. The patient was discharged from the hospital on the 22th day of hospitalization.

Discussion

Lemierre syndrome, otherwise known as postanginal sepsis or necrobacillosis, received its name from a comprehensive clinical description by Lemierre in 1936. This syndrome is a relatively uncommon entity that shows internal jugular vein thrombosis and septic emboli, and it occurs primarily in healthy young individuals; it usually follows an acute oropharyngeal infection caused by Fusobacterium necrophorum. This has been the most common pathogen found in Lemierre syndrome since Lemierre's original series (1) and it accounted for 81% of the cases in the recent review by Sinave and colleagues (2). Fusobacterium necrophorum is a common saprophyte of the oral cavity, but when it results in septicemia, a fulminant infection occurs that necessitates early and prolonged intravenous antibiotic therapy. The incidence of Lemierre syndrome has significantly decreased and the outcomes have improved with the wide spread use of antibiotics. Yet, despite the advances in chemotherapeutics, pulmonary embolism can occur in these patients, leading to serious clinical conditions.

Infection of the parapharyngeal space, which may occur secondary to the direct spread of an oropharyngeal infection or by lymphatic or tonsilar venous dissemination, is central to the development of Lemierre syndrome (3). Internal jugular venous thrombosis results from the adjacent inflammatory process or the extension from the tonsillar veins acts as a nidus of infection, and this can spread hematogenously and result in septicemia and septic embolization, which occur most commonly in the lungs (2). Another frequent complication of Lemierre syndrome is metastatic septic joint effusion and arthralgia. Hepatic and splenic abscess, osteomyelitis, meningitis, epidural abscess and diffuse encephalopathy have also been described in association with this syndrome (2 - 4). The presence of jugular venous thrombophlebitis is considered a hallmark of the illness, though it is not always present. Blood cultures should confirm an anaerobic septicemia caused by Fusobacterium, but this may be negative because of the use of antibiotics prior to hospitalization (5). In our case, no pathologic organism was isolated, and this was probably due to the prior antibiotic therapy. So, although any pathologic organism was not isolated, our case is compatible with Lemierre syndrome both clinically and radiologically.

Treatment of Lemierre syndrome is a prolonged course of the correct intravenous antibiotics that can cover the anaerobes. Anticoagulation therapy for the jugular vein thrombosis is sometimes used, and more intense therapies such as internal jugular vein ligation or surgical drainage of abscesses are occasionally necessary (6).

The radiologic findings of Lemierre syndrome are essential for its diagnosis. Awareness of this syndrome by the radiologist should lead to the suggestion of Lemierre syndrome whether by demonstration of the internal jugular venous thrombosis or through chest CT findings that are suggestive of septic emboli (3).

The critical feature of Lemierre syndrome is the pres-

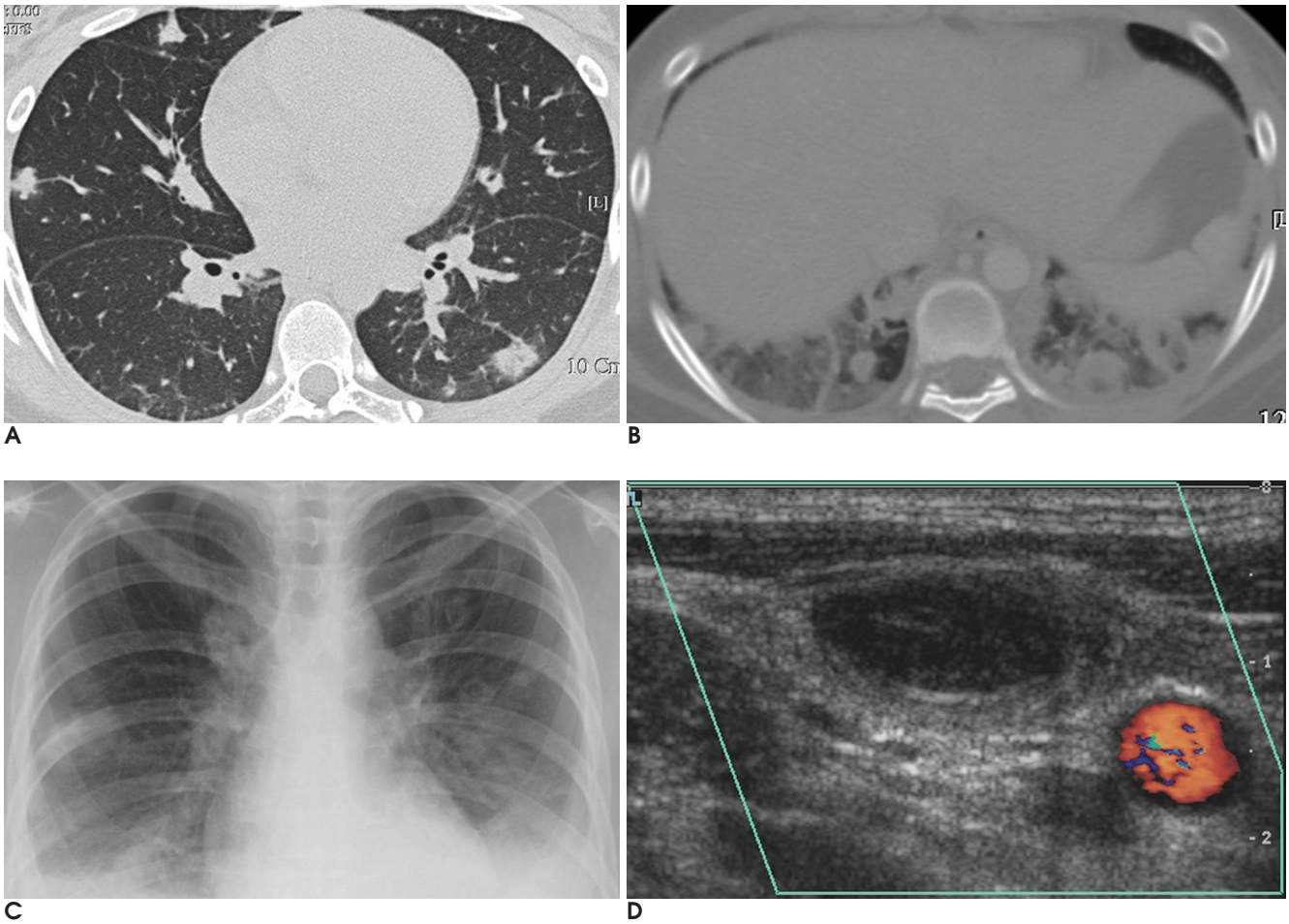

Fig. 1. A 16-year-old female with Lemierre syndrome
A. The HRCT scan obtained on the day of admission shows multiple bilateral peripheral nodules or consolidation with surrounding ground glass attenuations. Some of the lesions show connection with pulmonary vessels, and this suggests septic emboli.
B. The CT scan obtained on the 2nd day of hospitalization reveals septic emboli with cavitary changes in some lesions.
C. The chest radiograph obtained on the 7th day of hospitalization shows cavitary nodules in the left upper and right middle lung zones with bilateral pleural effusions.
D. Ultrasonography of the neck obtained on the 8th day of hospitalization shows non-compressible echogenic occlusive thrombus with venous wall thickening and segmental fusiform dilatation. Note the absence of flow in the right internal jugular vein. This finding is a hallmark of Lemierre syndrome.
E. The Neck CT scan performed on the 20th day of hospitalization shows right internal jugular vein thrombus (arrow).

ence of septic pulmonary emboli with the appearance of multiple peripheral, round, and wedge-shaped opacities that progress rapidly to cavitation (3). In our case, an overt feeding vessel leading into some of the nodules was noted and this so-called feeding vessel sign is also characteristic of septic embolism. Peripheral enhancement with central areas of reduced attenuation is seen following the intravenous administration of contrast medium; this enhancement is a characteristic feature of septic infarction (7, 8). Huang et al (7) and Kuhlman et al (8) obtained the same results: for the patients with septic pulmonary embolism, CT was the first diagnostic imaging modality to produce results that were suggestive of the diagnosis of septic pulmonary emboli in 46% and 33% of their patients, respectively. CT is superior to chest radiography for providing additional information and for indicating the septic emboli. Ultrasonography of the neck is a noninvasive and readily available imaging tool to demonstrate thrombi. The findings of ultrasonography include the presence of non-compressible thrombus, which are frequently associated with venous distension, and the absence of flow (9).

In conclusion, Lemierre syndrome is an uncommon malady, but it remains as a potentially life-threatening disease that requires a high index of suspicion for diagnosis. Early diagnosis and prompt treatment is essential to prevent the metastatic dissemination of septic emboli and to reduce the morbidity and mortality of this disease. The radiologist should be alert for this syndrome, particularly in the young populations, and they should suggest this diagnosis when the radiologic and clinical features, following an acute oropharyngeal infection, are compatible with septic emboli.

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르미에르 증후군은 젊은 성인에서 급성 구인두 감염 이후에 내경정맥 혈전과 패혈성 폐색전증을 특징으로 하는 드문 질환으로 이 질환에 대한 보고가 많지 않다. 저자들은 급성 편도선염을 앓고 있던 16세 여자환자에서 발생한 르미에르 증후군의 방사선학적 소견을 보고하고자 한다. 단순흉부촬영상 빠르게 진행하는 공동화를 보이는 결절들을 볼 수 있었고 HRCT에서는 패혈성 폐색전증을 시사하는 혈관과 연결된 다병소의 반점형 폐경화의 소견이 관찰되었다. 경부 초음파와 두부 CT 검사에서는 내경정맥 혈전이 보였다.