

## Papillon-Lefèvre syndrome with pyogenic liver abscess : case report focusing on radiological findings and review of the literature

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We present the results of CT and US in the rare case of Papillon-Lefèvre syndrome (PLS) with multiple pyogenic liver abscesses.

A 26-year-old woman presented with fever and right hypochondrial pain. At 3 years of age, she had been diagnosed as having PLS at another hospital. She had a history of abdominal, liver, and renal abscess. Again, a liver abscess was suspected, and *Staphylococcus aureus* was found in the drainage fluid specimen. She was transferred to our hospital. Upon admission, her temperature was 37.2°C and her pulse was 94 per minute. She had mild keratosis of the soles. Although she had a history of periodontitis, her permanent teeth were still in place probably because of continual treatment for periodontitis. The laboratory results showed a total white blood cell count of 12,500/ $\mu$ L and a C-reactive protein of 6.13 mg/dL. The nitroblue tetrazolium (NBT) reduction test showed a reduced response to latex stimulation (12%). Contrast-enhanced CT and US of the abdomen revealed multiple ill-defined hypodense and inhomogeneous masses with peripheral enhancement in the liver (Fig. 1). Our diagnosis was pyogenic liver abscess associated with PLS, as no other causes of liver abscess could be demonstrated. After treatment, the patient recovered and was discharged four months after admission. Further CTs performed 16 months later showed that the lesions were well defined and reduced, suggesting absorption of the liver abscess (Fig. 2). The patient is now symptom-free, and periodic observation in the outpatient department is maintained.

Although no gene test was performed in our case, the clinical diagnosis of PLS had been made at another hospital. The history of infections, keratosis of the soles and the low value of the NBT reduction test are compatible with the diagnosis of PLS. Hematogenous dissemination from the inflamed gingiva is suggested as pathogenetic mechanism of liver abscess associated with PLS (1,2). Therefore, pyogenic liver abscesses with PLS might be multiple, as in our case. The image findings of pyogenic liver abscess with PLS have not been discussed at all. There have been only two reports that showed contrast-enhanced CT (2,3). Anuradha *et al.* (3) reported that CT revealed multiple ill-defined, heterodense liver abscesses and multiple hypodense recurrent liver abscesses with few septations and peripheral enhancement two years

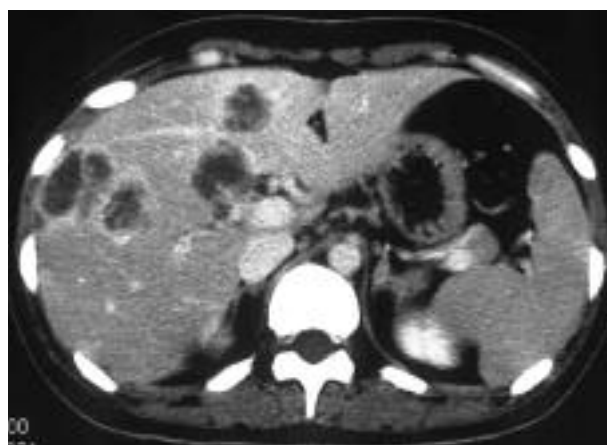


Fig. 1. — Contrast-enhanced CT on admission



Fig. 2. — Contrast-enhanced CT after 16 months

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later. Another report described a complex mass with hypoechoic and hyperechoic regions. However, no images were presented. Other reports describing pyogenic liver abscesses with PLS also showed neither images nor CT or US findings. In CT, common pyogenic liver abscesses appear as well-defined hypoattenuating lesions, unilocular with smooth margins or complex with internal septa, and having an irregular contour with faint rim enhancement (4). In US, their appearance ranged from hypoechoic to hyperechoic, with varying degrees of internal echoes (4). Hepatic abscess can vary according to stage of inflammation, and this may explain these variations. Thus, no differences of CT or US findings were observed between common pyogenic liver abscesses and those with PLS except in their number – multiple or solitary. The disease outcome of pyogenic liver abscess with PLS was not discussed at all. Three of eight reported patients had histories of liver abscess or recurrence (1,3). Our patient also had a history of liver abscess and required long hospitalization, but the lesions remained. Pyogenic liver abscess with PLS appears to have been

recurrent or to have taken a long time to heal because of immunodeficiency. PLS should be included in the differential diagnosis of liver abscess in young people, especially in multiple and prolonged or recurrent cases. Some cases in which liver abscess led to the discovery of PLS were even described (2,3).

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