PAPILLON LEFEVRE SYNDROME PRESENTING AS LIVER AND LUNG ABSCESS

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INTRODUCTION

We report two cases of Papillon Lefevre syndrome, which is a rare autosomal recessive condition. One patient presented with a staphylococcal liver abscess and the other with a lung abscess. Patients with this syndrome have altered immunity and are susceptible to infection, many of them develop liver abscess and the organism involved is usually staphylococcus Aureus. The salient features of this syndrome are hyperkeratosis of palms and soles, gingival inflammation with loss of teeth.

CASE REPORT

CASE 1

A young man of 20 years age was admitted to our unit in April 1989 with complaints of fever and pain right hypochondrium of one month duration.

He was the eighth child of the family born after normal pregnancy and delivery. When he was 4 years old, the skin of his palms and soles became red, thick, scaly and fissured. At birth the nails were normal but few days later, they began to show signs of deformation. The deciduous teeth erupted fully up to the age of 5 years, then inflammation of the periodontal tissues started and he lost all his teeth at the age of seven years. At the age of 15 years all his permanent teeth fell except 3 loose teeth in his mouth.

He has 6 brothers and 5 sisters. Two of his brothers, (35 & 23 years of age) have similar changes.

On examination he was thin and tall with the skin over palms, soles and medial malleoli reddish, moderately hyperkeratotic and scaly. Nails were deformed with opaque grey hue, elongated, thickened and flat with transverse ridges. Oral examination revealed recession of the gums and alveolar processes. Only three teeth were present. He was pale and had temperature of 102 F. Liver was enlarged 6 cm below the costal margin and was tender.

His haemoglobin was 11.4 gm% and TLC was 24000/ Cmm. Differential leucocyte count showed Neutrophils 85% Lymphocytes 14% and ESR was 103 mm in first hour. Ultrasound of the abdomen showed an abscess in the right lobe of the liver, which was drained and the pus grew staphylococcus aureus.

His response to conservative management was unsatisfactory so he had open drainage of the abscess by the surgeon followed by appropriate antibiotics.

Fig. 1. Photograph of patient No.1 showing hyperkeratosis of his palms and loss of teeth.

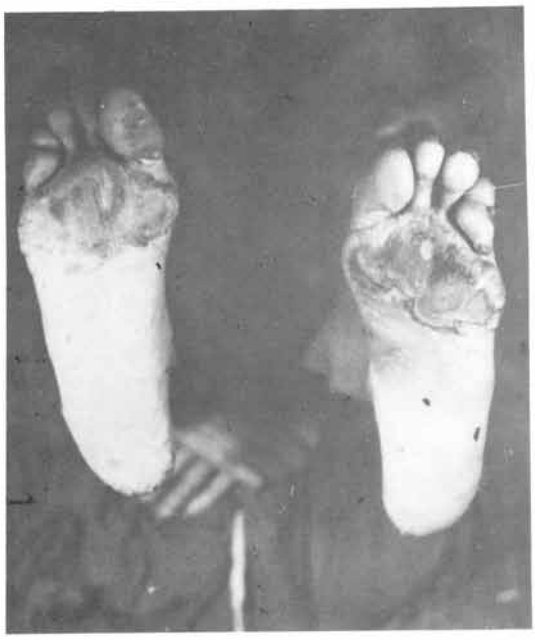

Fig. 2. Photograph of patient No.1 showing marked hyperkeratosis and scaling of his soles.

CASE 2

A 23 years old lady first presented to the out patient clinic with a history of cough productive of purulent sputum and dyspnoea of 3 months duration. On examination she was apyrexial. She was thin and tall and also had thickened scaly palms and soles and loss of teeth with only 5 teeth remaining in her mouth. Examination of the chest revealed inspiratory and expiratory wheezes and few crackles on the left midzone and base.

She is the first cousin of our first patient. None of her brothers and sisters had the above problems with their teeth and skin.

Investigations revealed high ESR, low haemoglobin and chest X-Ray showed an abscess in the left mid-zone. Sputum cultures grew strains of staphylococcus auras resistant to Ampicillin, Erythromycin and sensitive to Ofloxacin. (TARIVID).

She was admitted and commenced on Ofloxacin 200 mg orally twice daily, alongwith bronchodilators , and chest physiotherapy. She responded well and by the time she was discharged after 2 weeks she was clinically well and on the chest x-ray a small patch in left mid-zone remained. On follow up she remains well, and X-ray chest is back to normal.

DISCUSSION

Papillon Lefevre syndrome is a rare disorder of skin and periodontal supporting tissue first described by Papillon and Lefevre in 1942.[1] | Goftin et al[2] in a comprehensive review of 46 cases suggested that children displaying a familial disposition for the disorder demonstrated an autosomal recessive mode of , inheritance. They also estimated that the frequency of Papillon Lefevre syndrome is 1-4 per million persons in the general population, whereas, the carrier frequency may be 2-4 per thousand

population. Clinically the gingival condition appears soon after the deciduous teeth erupt and at the same time as the palmar hyperkeratosis appears.[3] By the age of 4 years, the deciduous teeth are normally lost. With the eruption of permanent teeth the process of periodontal destruction begins, again usually culminating in the early loss of all the teeth, although third molars do not seem to be affected.[4] Patients with this syndrome have increased susceptibility to infection due to immunological insufficiency and it may account for the progressive parodontopathia typical of Papillon Lefevre syndrome.[5] Staphylococcal liver abscess is not an uncommon occurrence in patients with this syndrome which is resistant to various antibiotics.

To our knowledge, a patient with Papillon Lefevre syndrome presenting with a lung abscess has not been described in the literature. This makes our patient No. 2 the first case of Papillon Lefevre syndrome presenting with a lung abscess. The possible mechanism apart from immunological insufficiency could be repeated gingival scpsis with aspiration resulting in lung abscess.

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