RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 7 | July 2014 | ISSN - 2249-555X

# A Rare Case of Papillon Lefevre Syndrome Associated with Primary Bronchiectasis

KEYWORDS: Papillon Lefevre Syndrome, Palmoplantarkeratoderma, bronchiectasis

| Dr. DIGUVINTI SREERAMULU | Dr. P.C.VASANTHA RAO | Dr. CHENNAKESAVULU DARA |
|---|---|---|
| M.D., ASSOCIATE PROFESSOR OF MEDICINE, GOVERNMENT GENERAL HOSPITAL, KURNOOL MEDICAL COLLEGE, KURNOOL, ANDHRA PRADESH. | M.D., ASSISTANT PROFESSOR OF MEDICINE, GOVERNMENT GENERAL HOSPITAL, KURNOOL MEDICAL COLLEGE,KURNOOL,. ANDHRA PRADESH. | JUNIOR RESIDENT IN GENERAL MEDICINE, GOVERNMENT GENERAL HOSPITAL, KURNOOL MEDICAL COLLEGE, KURNOOL, ANDHRA PRADESH. |

**ABSTRACT** *Papillon Lefevre Syndrome is a rare autosomal recessive syndrome that appears in childhood, characterized by diffuse transgredient Palmoplantarkeratoderma(PPKD) and aggressive periodontitis in both primary and secondary dentition with recurrent pyogenic infections. We report a rare case is associated with primary bronchiectasis which has not been reported so far to our knowledge.*

**INTRODUCTION:**
Papillon Lefevre Syndrome(LPS) is a rare autosomal recessive disorder characterized by palmoplantarkeratoderma(PPKD) and destructive periodontal disease affecting both the primary and secondary teeth [1,2].These patients may have decreased neutrophil,lymphocyte (or) monocyte functions and increased susceptibility to bacteria associated with recurrent pyogenic infections. Additional manifestations are nail dystrophy, hyperhydrosis and rarely pyogenic liver abscess [1,3]. The symptoms may worsen in winter and associated with painful fissures.

A genetic predisposition with greater frequency of occurrence in consanguineous offspring has been reported [4]. Here our case of PLS associated with primary bronchiectasis is rare.

**CASE REPORT:**
A 25 year old female patient presented to medical unit with chief complaints of fever,cough with expectoration and breathlessness. There was history of eczema over the both palms and soles. All deciduous teeth had exfoliated at early age, now she is edentulous. She was second child of family born of consanguineous marriage, her younger brother had similar dental and skin problems.

General examination she is tall and thin, her palms and soles shows symmetric, well defined keratotic plaques with fissures.(Fig-1,2,3) Nails were thickened, opaque, elongated, curved with transverse ridges. Oral examination reveals edentulous.(Fig-4) Respiratory system examination shows bilateral coarse leathery crepitations. Blood investigations shows rised ESR,CRP and elevated total leukocyte count. HRCT Chest shows bronchiectatic changes in the both the lung fields.(Fig-5) Histopathological examination of skin lesion shows compact hyperkeratosis, parakeratosis, hypergranulosis and dermis showed increased in fibrocollagenous tissue.(Fig-6)

After considering the clinical and histological features a diagnosis of Papillon Lefevre Syndrome was made.

**Fig-1: Palmar keratosis.**

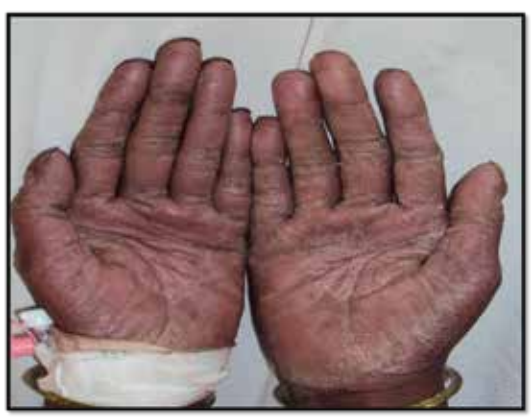
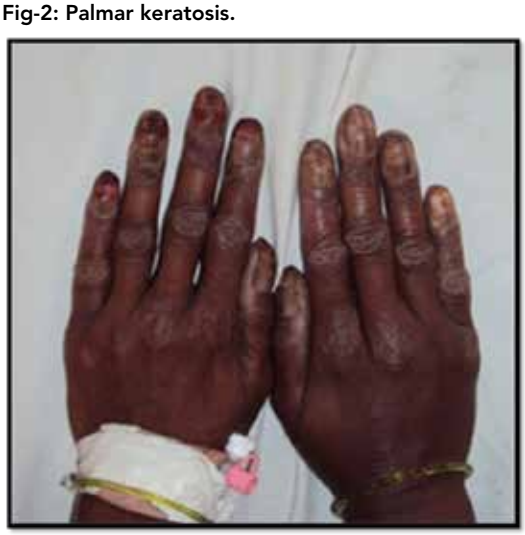


**Fig-2: Palmar keratosis.**



Volume : 4 | Issue : 7 | July 2014 | ISSN - 2249-555X

**Fig_3: Plantar keratosis**

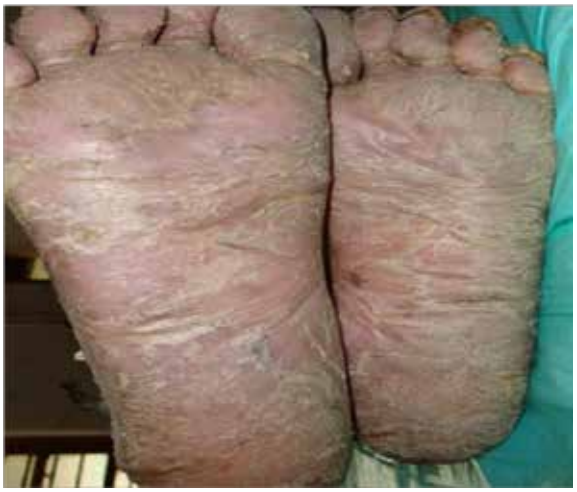


**Fig-4: Absence of teeth.**

**Fig-5: HRCT Chest shows bilateral bronchiectasis**

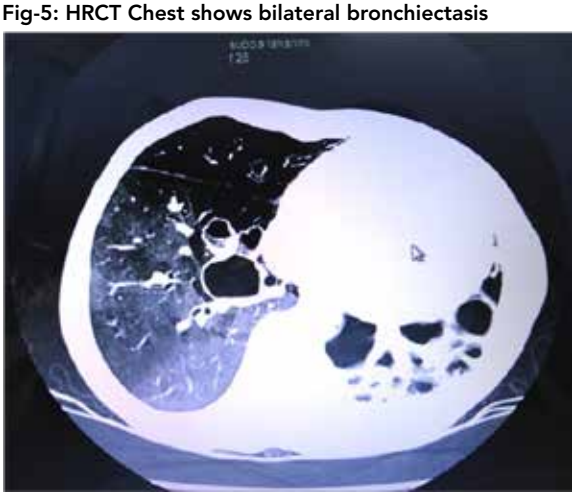
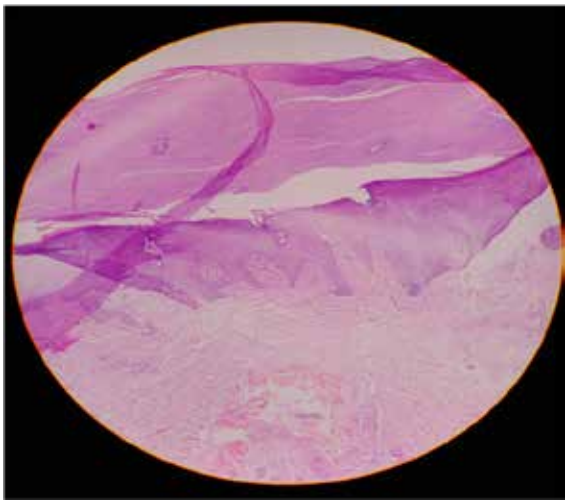


**Fig-6 Histology suggestive of keratosis:**



**HISTOLOGICAL EXAMINATION showing**

- compact Hyperkeratosis
- Parakerarosis
- Mild acanthosis.
- Hypergranulosis.
- Papillomatosis.
- Dermis showed inrease in fibrocollagenous tissue .

**DISCUSSION:**

Papillon Lefevre Syndrome characterized firstly by two French Physicians Papillon and Lefevre in 1924. It has a prevalence of 1-4 cases per million persons and both male and females are equally affected with no racial predominance [2,5,6]. With both parents are recessive carriers, there is a 25% chance of producing offspring with PLS. Various etiopathogenic factors are associated with the syndrome, but a recent report has suggested that the condition is linked to mutations of cathepsin-C gene located on chromosome 11q14 [7].

Haneke used the following the three criteria to classify a case as PLS [8]:

(1) Palmoplantar hyperkeratosis.
(2) Loss of primary and permanent teeth.
(3) Autosomal Recessive inheritance .

In the present case clinically and histologically suggestive of palmoplantarkeratoderma and edentulous diagnosed as PLS.

**CONCLUSION:**

Cases of PLS with lung and liver abscess were reported in the literature. To our knowledge a patient with PLS presenting with primary bronchiectasis has not been described in the literature, whether this association was just a coincidence or due to some underlyimg pathology is not clear.

**REFERENCE**

1. Bergman R, Friedman-Birnbaum R. Papillon-Lefèvresyndrome: a study of the long-term clinical course of recurrentpyogenic infections and the effects of etretinate treatment. | Br J Dermatol 1988;119:731–36. | | | 2. Subramaniam P, Mathew S, Gupta K K. Papillon-Lefevre Syndrome: A case report. J | exfoliation of all deciduous teeth were started Indian Soc Pedod Prev Dent 2008;26:171-4. | | 3 Khandpur S, Reddy BS. Papillon-Lefèvre syndrome withpyogenic hepatic abscess: a rare association. Pediatr Dermatol2000;18:45–7. | | | 4. Hattab FN and Amin WM (2005) Papillon Lefevre syndrome with albinism: A | review of Literature and report of 2 brothers. Oral Surg Oral Med Oral Pathol | Oral Radiol Endod 100: 709-16. | | | 5. Neville BW, Damm DD, Allen CM,Bouquot JE. Papillon-Lefevre Syndrome.Oral and Maxillofacial Pathology, 3rd edition. Noida, Elsevier Inc, 2009;176-8. | | 6. Nagaveni NB, Suma R, Shashikiran ND, SubbaReddy VVPapillonLefevresyndrome:Report of two cases in the same family. J Indian Soc Pedod Prevent Dent 2008;:78-81. | | 7.Ghaffer KA, Zahran FM, Fahmy HM, Brown RS. Papillon–Lefèvre syndrome. Neutrophil function in 15 cases from 4 families in Egypt. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology and Endodontics 1999; 88: 320– 325. | | 8.Haneke E. The Papillon–Lefèvre syndrome: keratosis palmoplantaris with periodontopathy. Human Genetics1979; 51: 1–35. |