



Papillon Lefe'vre Syndrome with Hepatic Abscess

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Abstract

PLS is a rare condition with an incidence of between one and four persons per million. The disease is an autosomal recessive inheritance and presents with palmoplantar keratoderma and destructive periodontitis usually beginning in early childhood [1].

Case Report

A 5-year-old boy was admitted to our dermatology department from the Pediatric Surgery Department of the Erciyes University. The patient presented hyperkeratosis on the palms, soles, and knees. He had these lesions for about four years. He admitted to Pediatric Surgery Department with stomachache and fever. His parents noticed that he had painful swelling of the gums and losing of his primary teeth. They also emphasized that he had skin abscess for three times. His medical history revealed no previous serious illness and he was of normal intelligence. His family history showed that he was born from a consanguineous marriage and his uncle also had palmoplantar hyperkeratosis. Dermatological examination showed bilateral, symmetrical hyperkeratotic plaques on the soles, palms, and knees (Figures 1, 2 and 3). He had a post-op drain on his hypocondrium. Oral examination showed that all the primary teeth were present except for four incisors. Oral hygiene was poor with significant plaque accumulation (Figure 4). His complete blood count and routine biochemical markers were within normal limits. Histopathologic examination of the hyperkeratotic plantar skin revealed hyperkeratosis, hypergranulosis, acanthosis and a dense perivascular lymphocytic infiltration. Differential diagnosis included an *Echinococcus granulosus* (hydatid cyst) and pyogenic liver abscess. Culture of hepatic abscess material yielded *S.aureus*. The bacteria identified by using conventional methods based on colony morphology on 5% blood agar, gram stain, catalase and coagulase tests [2].

He was treated with ornidazole, teicoplanin and amikacin intravenously for two weeks, after drainage of the abscess. The patient recovered dramatically. PLS diagnosis was made after dermatologic and dental examination. Of the possible differential diagnosis Unna-Thost was ruled out as he had periodontitis, Mal de Meleda was ruled out as he did not have extensor hyperkeratosis and Olmsted was ruled out as he did not have an eccrine gland dysfunction. L3Vaseline-Salicylic acid (20%) was prescribed for the hyperkeratotic lesions.

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Figure 1: Bilateral, symmetrical hyperkeratotic plaques on the soles.



Figure 2: Bilateral, symmetrical hyperkeratotic plaques on the knees.

Discussion

PLS is a rare disease with an incidence of between one and four persons per million [3]. The disease has an autosomal recessive inheritance and presents with palmoplantar keratoderma and destructive periodontitis usually beginning in early childhood [1]. In literature late onset cases are reported [4]. Our patient had a typical palmoplantar keratoderma. CTSC encodes the cathepsin C protein, which is a member of the peptidase C1 Family [5]. Studies showed the mutations of the cathepsin-C gene in PLS, located on chromosome 11q14.1-q14.3 [6,7]. The cathepsin-C gene is expressed in epithelial regions i.e. soles, palms, knees, and keratinized oral gingiva, and in many immune cells i.e. macrophages, polymorphonuclear leukocytes. The other conditions related with the mutation of the cathepsin C gene are prepubertal periodontitis and Haim-Munk syndrome. The



Figure 3: Bilateral, symmetrical hyperkeratotic plaques on the palms.



Figure 4: Poor oral hygiene with significant plaque accumulation.

common manifestation of all these three syndromes is severe early-onset periodontitis. Haim-Munk syndrome has been described as an autosomal-recessive genodermatosis characterized by progressive early-onset periodontitis and congenital palmoplantar keratoderma. It also exhibits atrophy of nails, arachnodactyly, acroosteolysis, and deformity of the phalanges in the hands [8]. The other conditions that can be included in the differential diagnosis are Greither syndrome, Howel-Evans syndrome and keratosis punctata. Even though all these diseases are associated with palmoplantar hyperkeratosis, periodontopathy is not seen in them [9]. Our patient showed classic events of gingivitis, periodontitis and loss of teeth.

Neutrophil-function test in PLS showed reduced response to *Staphylococcus spp.* and *A. actinomycetemcomitans* [10] and microbiological studies showed that *Actinobacillus actinomycetemcomitans* has a major role in the periodontal pathogenesis in patients with PLS. Other pathogens have also been reported, including *Prevotella nigrescens*, *Fusobacterium nucleatum* and *Peptostreptococcus micros*, *Eikenella corrodens*, *Porphyromonas gingivalis*, *Treponema denticola*, *Porphyromonas gingivalis*, *Bacteriodes forsythus*, and *Prevotella Intermedi* as well as *Cytomegalovirus* and *Epstein-Barr type 1 virus* [11]. A rare feature of PLS that our

patient had, is hepatic abscesses. Pyogenic liver abscess usually originates from the seeding of the liver by pathogenic bacteria through a hematogenous route. The most common etiologic agent is *S aureus*; as our patient. To date, there are only a few case reports of PLS in the literature those were complicated with pyogenic liver abscess [12,13]. Another feature of PLS may be intracranial calcification. We did not detect any calcification on his cranial computed tomography.

Conclusion

we have described a 5-year-old Turkish boy diagnosed as PLS who exhibited palmoplantar keratoderma, periodontitis, skin involvement and a rare feature; hepatic abscesses.

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