OLGU SUNUMU CASE REPORT

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Prurigo Pigmentosa: A Report of Two Patients in Possible Association with Essential Fatty Acid Deficiency

Prurigo Pigmentoza: Muhtemel Esansiyel Yağ Asidi Eksikliği ile İlişkili İki Olgu Sunumu

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One of that cases was presented as a poster in Dermatopatology & Esthetics Academy 2018 Congress 8-11 March, Ankara, Turkey.

ABSTRACT Prurigo pigmentosa is a rarely seen dermatosis with pruritic reticular erythematous lesions generally located in the midline of the trunk. Its etiology has not yet been fully understood. The lesions were clinically compatible with prurigo pigmentosa in two male patients aged 19 and 22, who were admitted to our outpatient clinic with brown-red itchy spots on their bodies, and both were on a low-carbohydrate and low-fat diet. They were completely cured by dietary change and tetracycline treatment. A low-carbohydrate diet or ketosis is commonly seen with prurigo pigmentosa and the disease may be regressed by diet changes. Ketone bodies are accused in the etiology of prurigo pigmentosa but the disease is not seen in each patient with ketosis. Low carbohydrate intake or ketosis affects fatty acid metabolism. We believe that changes in fatty acid amounts or composition in the skin may probably play a role in the etiopathogenesis of prurigo pigmentosa.

Keywords: Prurigo pigmentosa; ketogenic diet; essential fatty acids; keto rash

ÖZET Prurigo pigmentosa, genellikle gövde orta hattında yerleşen pruritik retiküler eritematöz lezyonlarla karakterize nadir görülen bir dermatozdur. Etiyolojisi henüz tam olarak anlaşılamamıştır. Polikliniğimize vücudunda kahverengi-kırmızı kaşıntılı lekeler şikâyeti ile başvuran 19 ve 22 yaşındaki 2 erkek hastada lezyonlar klinik olarak prurigo pigmentosa ile uyumlu idi ve her ikisi de karbonhidrat ve yağdan fakir diyet uygulamaktaydı. Diyet düzenlenmesi ve doksisiklin tedavisi ile olguların lezyonları tamamen düzeldi. Düşük karbonhidratlı diyet veya ketozis prurigo pigmentozaya sıklıkla eşlik eder ve hastalık kişilerin eski diyete geri dönmeleri ile iyileşir. Keton cisimleri prurigo pigmentoza etiyolojisinde suçlanmaktadır, ancak ketozis olan her hastada hastalık görülmemektedir. Düşük karbonhidrat alımı ile birlikte yağ alımı da sınırlandırıldığında göreceli olarak yağ asitlerinin ketozise bağlı kullanımlarının artışı ve yerine konamaması prurigo pigmentoza etiyopatogenezinde muhtemel rol oynuyor olabilir.

Anahtar Kelimeler: Prurigo pigmentoza; ketojenik diyet; esansiyel yağ asitleri; keto döküntüsü

Numerous cases of prurigo pigmentosa associated with ketosis and ketogenic diet have been reported in the literature.[1] We aimed to present these cases to draw attention to the importance of diet and metabolism in the etiopathogenesis of prurigo pigmentosa. We present two cases of prurigo pigmentosa who used ketogenic diet and used a fat burner in addition to a low-carbohydrate and fat-restricted diet.

CASE REPORT 1

A 19-year-old male patient with skin type 1 was admitted to our clinic with the complaint of an itchy rash on the chest and back for 2 weeks. On dermatologic examination, erythematous papular lesions that tend to coalesce on the chest and reticular hyperpigmented macules were seen on the upper back. The clinical diagnosis was consistent with prurigo pig-

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mentosa (Figure 1). It was determined that the patient had been on a ketogenic diet for 2 months, but also limited the consumption of fat. He had no accompanying disease and a history of drug use. The patient returned to his normal diet, and doxycycline therapy was given 200 mg/day for 2 weeks. Lesions disappeared within 3 weeks.

CASE REPORT 2

A 22-year-old male patient with skin type 1 was admitted to our clinic with the complaint of an itchy rash on the chest area for 10 days. Reticular erythematous macules and papular lesions that tend to coalesce covering the middle of the chest were seen on dermatologic examination. Clinical features were consistent with the diagnosis of prurigo pigmentosa (Figure 2). He was taking a thermogenic fat burner supplement containing green tea, citrus, and red pepper extract, caffeine, phenylalanine, and unroasted coffee beans, as well as limiting daily calorie and fat intake. The fat-burning food supplement used by the patient was discontinued and he was reassured to switch to his normal diet. The lesions completely regressed within 4 weeks. The consent form was taken from our patients.

DISCUSSION

Prurigo pigmentosa is a rare skin disease with an unknown etiology. Although a few cases have not been associated with diet and systemic disease, there are many cases of prurigo pigmentosa reported in the literature, which have been reported to develop in conditions that cause ketosis, such as a ketogenic diet or diabetic ketoacidosis.[2] In cases of prurigo pigmentosa due to ketosis, skin lesions also improve when ketosis improves.[1]

The basic principle in the ketogenic diet is to compensate for the energy deficit that has arisen by increasing the consumption of fat and protein while severely restricting carbohydrate intake. In this situation, fatty acids are used for energy production and break down into ketone bodies.[3]

Some people limit their fat intake during the ketogenic diet as in our cases. Our first case followed a ketogenic diet, but also restricted fat intake. Our second case was using a food supplement had a fat-burning feature, but also restricted carbohydrate and fat consumption. An acute possible essential fatty acid (EFA) deficiency may occur during the ketogenic and fat-free diet, both due to the increased use of EFAs to compensate for the energy deficit and their inadequate intake. Therefore, EFA deficiency along with ketosis may play an additional factor in the etiology of prurigo pigmentosa.

A study in rats supports the view that the main changes in the fatty acid composition of mice on a

FIGURE 1: Patient 1, on ketogenic diet, reticular erythematous papular lesions that tend to coalesce on the chest and upper back.

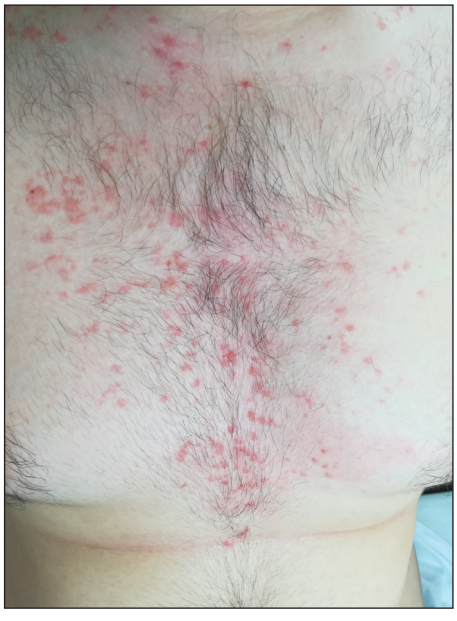

FIGURE 2: Patient 2, with the use of thermogenic fat burner, erythematous reticular papular lesions covering the the chest area.

ketogenic diet are due to the mobilization of EFA group polyunsaturated fatty acids from adipose tissue to the liver and brain. Fatty acids are released from adipose tissue for energy production during the ketogenic diet.[4] In ketosis, ketone bodies produced from fatty acids accumulate in tissues and lead to cytotoxic effects and inflammation through intercellular adhesion molecule 1.[1]

However, the disease does not develop in every patient with ketosis. This may be related to the severity of ketosis, or it may be due to the limitation of fat intake during the ketogenic diet. It may result in an acute fatty acid deficiency due to their inadequate fat intake and their increasing use to compensate for the current energy deficit. Fatty acid deficiency may play a role in the pathogenesis of the disease along with ketosis.

Prurigo pigmentosa can also be seen in malabsorption cases leading to nutritional deficiency such as bariatric surgery and hyperemesis.[5] In cases of liver failure, malnutrition and malabsorption can develop.[6] A skin disease similar to prurigo pigmentosa has been reported in a patient with dermatitis herpetiformis accompanying celiac disease.[7] In such diseases that can lead to malabsorption or malnutrition, the development of prurigo pigmentosa may occur due to poor absorption or inadequate intake of EFA in addition to ketosis.

Essential fatty acids are a group of polyunsaturated fatty acids and consist of a group of omega 3 and omega 6 fatty acids. The best known alpha-linoleic acid from the omega 3 group is linoleic acid from the omega 6 group. EFA plays an important role in skin barrier functions. They also have anti-inflammatory, photoprotective, and supportive properties in wound healing.[8,9] In cases of EFA deficiency in humans, a dermatitis-like picture appears, characterized by erythema and dryness.[10,11]

Prurigo pigmentosa can be seen more frequently in light-skinned people of the Caucasian race. After Japan and Korea, most cases have been reported in Turkish society. There are also cases reported from Italy, Sicily, China, and the USA.[2] The development of prurigo pigmentosa may have been getting easy because our patients have Caucasian skin type.

The prevalence of prurigo pigmentosa in Japanese society may also be related to the eating habits of this community, apart from its racial characteristics. In a study that examines the diet of the Japanese community, it is reported that about a third of young girls in the age group 20 receive less than 80% of their daily energy needs. However, it was determined that the oil consumption of the population examined in this study was not too low.[12] In this case, even if the fat consumption is normal, insufficient calorie intake within the diet may play a role in the etiology of prurigo pigmentosa secondary to genetically predisposition in light-skinned individuals by ketosis regardless of fatty acid deficiency.

The effectiveness of tetracyclines in the treatment of prurigo pigmentosa may be due to the non-antibiotic effects of these agents. Tetracyclines have antioxidant properties in addition to their effects on cytokine regulation, chemotaxis, and activation of leukocytes.[13] Tetracyclines are also effective in fatty acid metabolism. Tetracyclines have been shown to have anti-lipolytic effects on fat cells.[14] In another study, oral tetracycline hydrochloride for acne treatment has been shown to significantly reduce the average free fatty acid/triglyceride ratio in hair lipids.[15] The therapeutic efficacy of tetracyclines in prurigo pigmentosa may be related to its effects on lipid metabolism in addition to its other features.

Changes in the amount or composition of fatty acids in the tissue may play a role in the etiology of prurigo pigmentosa. It may be possible that the development of prurigo pigmentosa in younger age groups and people with low body fat ratio may be associated with a ketogenic or low-carbohydrate diet, but prurigo pigmentosa may also develop due to metabolic diseases that may lead to ketosis.

Hence we suggest a possible malnutrition or absorption disorder which can lead to ketosis should be investigated and the daily calorie values and fat consumption should be questioned in every patient with prurigo pigmentosa.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Semih Güder;* ***Design:*** *Semih Güder;* ***Control/Supervision:*** *Hüsna Güder;* ***Data Collection and/or Processing:*** *Semih Güder;* ***Analysis and/or Interpretation:*** *Hüsna Güder;* ***Literature Review:*** *Semih Güder;* ***Writing the Article:*** *Semih Güder, Hüsna Güder;* ***Critical Review:*** *Hüsna Güder.*

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