CHAPTER 34

PALMOPLANTAR KERATODERMA (Q82.8)

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REMEMBER

- Palmoplantar keratoderma (PPK) is defined as the thickening of palmar and plantar areas without friction.
- Palmoplantar keratoderma has acquired and inherited forms. Hereditary PPKs tend to occur at earlier ages of life.
- Some diseases such as pachyonychia congenita, Mal de Meleda, Olmsted, Papillon-Lefévre, KID (keratitis, ichthyosis, deafness), Naegeli-Franceschetti-Jadassohn, Clouston, Huriez, Naxos, Haim-Munk, and KLICK syndromes are among the causes of hereditary PPK.
- Hereditary PPKs have autosomal recessive and dominant, mitrochondrial and X-linked forms.
- Clinical evaluation of PPK patients begins with the division of cases into hereditary or acquired forms.
- Cases of hereditary PPK are first evaluated by features of dissemination (diffused, focal or point), presence/absence of transgressions, accompanying symptoms and, of course, family history.
- It is important that hereditary cases are evaluated together by dermatologists and medical geneticists.
- Various diagnostic tests, including complete blood count, fungal scrapings, chest X-ray, thyrotropin, antinuclear antibodies (ANA), rapid plasma reagent for syphilis (RPR), purified protein

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Orthopedics/Plastic Surgery

- The patients suffering from PPK should be consulted for the treatment of pseudoainhums, and/or scleroatrophies.

Infectious Diseases

- Systemic pyogenic infections should be detected.
- The relationship of acquired PPK with crusted scabies, tuberculosis, leprosy, human papilloma virus (HPV), syphilis and HIV has been described. In case of doubt, an evaluation is required.

Dentistry

- In hereditary cases, periodontal, and dental disorders will both give an idea about the type of PPK and possible treatments will be planned if accessible.

Medical Oncology/Hematology

- If in doubt, PPK patients should be evaluated for esophageal and lung carcinomas.
- Female patients should also be screened for cancer of the breast, uterus, cervix and ovary.
- In addition, the relationship of acquired ichthyosis with Hodgkin's disease, non-Hodgkin lymphoma, some leukemias, multiple myeloma, Kaposi's sarcoma, and leimyosarcoma is known. The patients should be evaluated in terms of these diseases in case of any symptoms, and/or doubt.

Ophthalmology

- Patients should be evaluated by an ophthalmologist in terms of corneal inflammation, and ulceration.

Psychiatry

- Patients should be evaluated by a psychiatrist to determine probable intellectual disorders, anxiety, and depression.

Medical Genetics

- In hereditary cases, the related genetic tests should be requested when necessary.
- SLURP1 mutations, GJB2 connexin gene mutation, loricrin mutations, TRPV3 and PERP (TP53 apoptosis effector) defects can be examined.

Clinical Pharmacology

- It has been shown that various drugs may trigger PPK. Lithium, hydroxyurea, quinacrine, and venlafaxine are some examples. In case of doubt, patients should be evaluated together with a clinical pharmacologist.

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BÖLÜM 35

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HATIRLA

- Palmoplantar keratoderma (PPK) palmar ve plantar alanların sürtünme olmadan kalınlaşması olarak tariflenir.
- Palmoplantar keratoderma akkiz (=edinsel) ve kalıtımsal olarak ortaya çıkabilir. Kalıtımsal PPK'lar daha erken yaşlarda ortaya çıkar.
- Pakionişi konjenita, Mal de Meleda, Olmsted, Papillon-Lefévre, KID sendromu, Naegeli-Francheschetti-Jadassohn, Clouston, Huriez, Naxos, Haim-Munk ve KLICK sendromları gibi bazı hastalıklar herediter PPK nedenleri arasındadır.
- Herediter PPK'ların otozomal resesif ve dominant, mitrokondriyal ve X'e bağlı kalıtılan formları vardır.
- Klinik değerlendirme, vakaların kalıtsal veya edinilmiş olarak ayrılması ile başlar.
- Kalıtsal PPK vakaları ilk olarak yayılım (yaygın, odaksal veya noktasal), transgredienlerin varlığı/yokluğu, eşlik eden semptomlar ve elbette aile öyküsünün özellikleri ile değerlendirilir.
- Kalıtsal vakaların dermatolog ve tıbbi genetik uzmanınları tarafından beraber değerlendirilmesi önemlidir.

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Kadın Hastalıkları ve Doğum

- Klimakterium/menopoz dönemindeki kadınlarda akkiz PPK gelişebilir. Kadın PPK'lı hastalar şüphe durumunda değerlendirilmelidir. Bu keratoderma tipi Haxthausen hastalığı olarak da bilinir. Hastalar bu açıdan jinekolojik olarak incelenmelidir.

Ortopedi/Plastik Cerrahi

- Psödoainhumların ve/veya skleroatrofilerin tedavisi açısından danışılmalıdır.

Enfeksiyon Hastalıkları

- Sistemik piyojenik enfeksiyonların tespiti yapılmalıdır.
- Akkiz PPK'nın krutlu skabiyez, tüberküloz, lepra, insan papilloma virüsü (HPV), sifiliz ve HIV ile ilişkisi tanımlanmıştır. Şüphe durumunda mutlaka değerlendirme yapılmalalıdır.

Diş Hastalıkları

- Herediter vakalarda peridontal hastalıklar ve diş bozuklukları hem PPK'nın tipine dair fikir verecektir hem de olası tedaviler mümkünse planlanacaktır.

Medikal Onkoloji/Hematoloji

- Eğer şüphe varsa hastalar özofageal ve akciğer karsinomları açısından mutlaka değerlendirilmelidir.
- Kadın hastalar ayrıca meme, uterus, serviks ve over kanseri açısından taranmalıdır.
- Ayrıca edinsel iktiyozun Hodgkin hastalığı, Hodgkin dışı lenfomalar, bazı lösemiler, multipl myelom, Kaposi sarkomu ve leimyosarkom ile ilişkisi bilinmektedir. Gerekli durumlarda bu açılardan da değerlendirme yapılmalıdır.

Göz Hastalıkları

- Korneal inflamasyon, ülserasyon açısından hastalar göz hastalıkları uzmanı tarafından değerlendirilmelidir.

Psikiyatri

- Olası entellektüel bozukluklar, anksiyete ve depresyon açısından hastalar psikiyatrist tarafından değerlendirilmelidir.

Tıbbi Genetik

- Kalıtsal vakalarda ilgili genetik testler istenmelidir.
- SLURP1 mutasyonları, GJB2 konneksin gen mutasyonu, loricrin mutasyonları, TRPV3 ve PERP (TP53 apopitozis effektörü) defektleri incelenebilir.

Klinik Farmakoloji

- Literatürde bazı ilaçların PPK'yı tetikleyebildiği gösterilmiştir. Lityum, hidroksiüre, kinakrin ve venlafaksin bunlardan bazılarıdır. Şüphe durumunda hastalar klinik farmakolog ile birlikte değerlendirilmelidir.

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