

CHAPTER 27

LINEAR IgA DISEASE (L13.8)

Ozan YILDIRIM¹
Zekayi KUTLUBAY²

REMEMBER

- ▶ Linear IgA bullous dermatosis (LABD) (=linear IgA disease) is a rare immune-mediated disease seen in both children and adults. The pediatric form is named as chronic bullous disease of childhood.
- ▶ The overall incidence of linear IgA disease (LAD) in Western Europe is less than 0.5 per million, and is slightly predominant in women. The global prevalence is not fully known.
- ▶ LAD has a genetic autoimmune susceptibility and the disease has been associated with HLA DR2, and DR3 loci.
- ▶ It has been reported that LAD can be triggered by bacterial/viral diseases or drugs.
- ▶ The disease is defined by the presence of a homogeneous accumulation of IgA in a linear band at the dermal-epidermal junction (DEB). Target antigens may be LAD antigen 97/120, BP 230, BP 180 or type VII collagen.
- ▶ There are three main types of LAD: drug-induced, pediatric and adult forms.
- ▶ Localized, morbiliform, erythema multiforme-like, targetoid lesions and Stevens-Johnson-like presentation have also been described.
- ▶ Classically, in adults, LAD presents as diffuse tense vesicles that are aligned in arcuate or ring-shaped (called strings of pearls, crowns of jewels, or rosette appearance).

¹ MD., Basaksehir Cam and Sakura City Hospital, Dermatology Clinic, yyildirimoza@gmail.com

² MD., Istanbul University-Cerrahpasa/Cerrahpasa School of Medicine, Department of Dermatology, zekayikutlubay@hotmail.com



Otorhinolaryngology

- ▶ In cases of severe airway involvement, the patient should be evaluated for the development of airway obstruction, and strictures.

Gastroenterology

- ▶ In patients with linear IgA disease, esophageal involvement should be evaluated and followed up for prevention stenosis.
- ▶ The relationship between inflammatory bowel diseases, especially ulcerative colitis, and LAD has been demonstrated. According to the patient's history, if necessary, evaluations should be performed in this respect.

Dentistry

- ▶ In terms of desquamative gingivitis, patients can be consulted with the dentist.

Medical Oncology

- ▶ Linear IgA disease may present as a paraneoplastic dermatosis in adults. The relationship between underlying malignancy and LAD is not clear, but the relationship has been reported in many cases in the literature.
- ▶ In case of suspicion of solid organ malignancy, a medical oncologist should be evaluated in this regard (e.g., prostate, colon, breast cancer, uterine carcinoma, esophageal cancer and renal cell carcinoma).

Hematology

- ▶ Associations with linear IgA dermatosis and adult multiple myeloma, Hodgkin's disease, non-Hodgkin lymphomas, and acute and chronic leukemias have been reported

in some cases. In case of doubt, the patient should be evaluated by a hematologist.

Ophthalmology

- ▶ Ocular involvement may occur in approximately half of the cases, and the possibility of involvement should be assessed in every patient. Patients should be followed up for ectropion, subconjunctival fibrosis, symblepharon, and corneal damage/blindness.

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LİNEER IgA DERMATOZU (L13.8)

Ozan YILDIRIM¹
Zekayi KUTLUBAY²

HATIRLA

- ▶ Lineer IgA büllöz dermatozu (LABD) (=Lineer IgA dermatozu) hem çocuklarda hem de yetişkinlerde görülen nadir bir immün aracılı büllü hastalıktır. Pediyatrik formu çocukluk çağıının kronik büllöz hastalığı olarak isimlendirilir
- ▶ Batı Avrupa’da genel lineer IgA dermatozunun (LAD) insidansı milyonda 0.5’ten azdır ve kadınlarda görülme olasılığı hafif olarak daha yüksektir. Global prevalans tam olarak bilinmemektedir.
- ▶ LAD’nin genetik bir otoimmün duyarlılığı vardır ve hastalık HLA DR2 ve DR3 gen lokusları ile ilişkilendirilmiştir.
- ▶ LAD’nin, bakteriyel/viral hastalıklar veya ilaçlarla tetiklenebileceği bildirilmiştir.
- ▶ Hastalık, dermal- epidermal bileşkede (DEB) lineer bir bant şeklinde homojen IgA birikiminin varlığı ile tanımlanır. Hedef antijenler LAD antigen 97/120, BP (büllöz pemfigoid) 230, BP 180 veya tip VII kollajen olabilir.
- ▶ Üç ana LAD türü vardır: ilaca bağlı, pediatrik ve yetişkin formlar.
- ▶ Lokalize, morbiliform, eritema multiforme benzeri, targetoid (=hedef benzeri) lezyonlar ile Stevens-Johnson benzeri durumlar da tarif edilmiştir.

¹ Dr., TC Sağlık Bakanlığı Çam ve Sakura Şehir Hastanesi, Dermatoloji Kliniği, yyildirimozan@gmail.com

² Dr., İstanbul Üniversitesi Cerrahpaşa, Tıp Fakültesi, Dermatoloji AD., zekayikutlubay@hotmail.com



Kulak Burun Boğaz Hastalıkları

- ▶ Ağır hava yolu tutulumu olan vakalarda hava yolu tıkanıklığı ve darlıkları açısından hasta değerlendirmelidir.

Gastroenteroloji

- ▶ LAD'li hastalarda yemek borusu tutulumu stenozun engellenmesi amacıyla değerlendirilmeli ve takip edilmelidir.
- ▶ Özellikle ülseratif kolit olmak üzere inflamatuvar bağırsak hastalıkları ile lineer IgA dermatozu arasındaki ilişki ortaya konulmuştur. Hasta hikayesine göre gerekirse bu açıdan değerlendirmeler yapılmalıdır.

Diş Hastalıkları

- ▶ Deskuamatif jinjivit açısından hastalar diş hekimine danışılabilir.

Medikal Onkoloji

- ▶ Lineer IgA hastalığı yetişkinlerde paraneoplastik bir dermatoz olarak karşımıza çıkabilir. Altta yatabilecek malignite ve LAD arasındaki ilişki net değildir ancak literatürde birçok vakada ilişki bildirilmiştir.
- ▶ Solid organ malignitesi şüphesinde bu açıdan medikal onkoloji uzmanınca değerlendirme yapılmalıdır (ör: prostat, kolon, meme kanseri, uterus karsinomu, özofagus kanseri ve renal hücreli karsinom gibi)

Hematoloji

- ▶ Lineer IgA dermatozu ile yetişkin multiple myelom, Hodgkin hastalığı, Hodgkin dışı lenfomalar ile akut ve kronik lösemiler ile ilişki bazı vakalarda bildirilmiştir. Şüphe varlığında hematoloji uzmanı tarafından değerlendirilme yapılmalıdır.

Göz Hastalıkları

- ▶ Oküler tutulum vakaların yaklaşık yarısında ortaya çıkabilir ve tutulum olasılığı her hastada mutlaka değerlendirilmelidir. Ektropion, subkonjunktival fibroz, simblefaron ve kornea hasarı/körlüğü açısından hastalar takip edilmelidir.

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