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PATIENT REPORT

Patient Age/Sex: 37 years Male

Specimen Collected: 21-Jun-22 13:50

Epithelial BMZ Ab, IgA **|Received: 22-Jun-22 09:45** **Report/Verified: 22-Jun-22 13:45**

Procedure	Result	Units	Reference Interval
Epithelial BMZ Ab,IgA	See Note [f1]		

Result Footnote

f1: Epithelial BMZ Ab, IgA
CLINICAL INFORMATION
Patient has pruritus with vesicles and bullae. Presumptive diagnosis is dermatitis herpetiformis versus epidermolysis bullosa acquisita versus bullous pemphigoid.

Specimen Details
S22-IP0000498 - Serum; Collected: 6/21/2022; Received: 6/22/2022

DIAGNOSTIC INTERPRETATION

Positive serum IgA basement membrane zone antibodies, consistent with linear IgA disease

(See Results and Comments)

RESULTS
Indirect Immunofluorescence (IIF)

Basement Membrane Zone (BMZ) IgA Antibodies

IgA: Positive, titer 1:2,560 (H), monkey esophagus substrate
Positive, epidermal pattern (roof), titer 1:1,280 (H), human split skin substrate

Reference Range:
Negative - Titer less than 1:10
Borderline - Titer 1:10
Positive (H) - Titer greater than 1:10

Localization Pattern on Human BMZ Split Skin:
Epidermal (roof), combined epidermal-dermal (roof and floor), or, dermal (floor) IgA BMZ antibodies = linear IgA disease (including linear IgA bullous dermatosis and chronic bullous disease of childhood)

IgA stronger than IgG epidermal (roof) BMZ antibodies = also possible linear IgA-predominant mucous membrane pemphigoid

(H) = high/positive

COMMENTS

Specific

*=Abnormal, #=Corrected, C=Critical, f=Result Footnote, H-High, i-Test Information, L-Low, t-Interpretive Text, @=Performing lab

Result Footnote

f1: Epithelial BMZ Ab, IgA

The positive IgA basement membrane zone antibodies reactive with monkey esophagus substrate and with human split skin substrate (also known as salt split skin) in an epidermal pattern by indirect immunofluorescence support the diagnosis of linear IgA disease.

Positive IgA basement membrane zone antibodies with epidermal localization on split skin substrate also can be:
- Co-expressed with IgG basement membrane zone antibodies in pemphigoid;
- Part of the characteristic findings in linear IgA/IgG bullous dermatosis;
- Observed in other autoantibody-associated diseases, including linear IgA variant mucous membrane pemphigoid or lupus erythematosus; or
- Nonspecific (generally, in low titer).

The presence of two antibody classes, IgA and IgG, with reactivity toward basement membrane zone may have implications for disease severity and treatment considerations. If indicated, further testing can be performed on this specimen for IgG basement membrane zone antibodies by contacting ARUP Client Services, 1-800-242-2787, option 2, with add-on test request(s) for:
- Basement Membrane Zone (Epithelial) Antibodies, IgG by IIF (ARUP test number 0092056),
- Bullous Pemphigoid (BP180 and BP230) Antibodies, IgG by ELISA (ARUP test number 0092566),
- Collagen Type VII Antibody, IgG by ELISA (ARUP test number 2010905).

Detection, levels, and patterns of diagnostic antibodies may fluctuate with disease manifestations. Clinical correlation is needed, including with direct immunofluorescence findings on a biopsy specimen and treatment status, with consideration for monitoring serum antibody profiles and levels to aid in assessing disease expression and activity, particularly for persisting, progressing, or changing disease, and in response to therapy.

General

Positive serum IgA epithelial basement membrane zone antibodies by indirect immunofluorescence are highly specific diagnostic markers for linear IgA disease and are present in sera of up to 80 percent of patients with linear IgA bullous dermatosis and chronic bullous disease of childhood. Linear IgA disease may be drug-induced, most commonly with vancomycin. IgA basement membrane zone antibodies also may be found in variant presentations of mucous membrane pemphigoid and epidermolysis bullosa acquisita. IgA basement membrane zone antibodies may be co-expressed with IgG basement membrane zone antibodies in some patients with pemphigoid, including mucous membrane/cicatricial pemphigoid, and develop in linear IgA/IgG bullous dermatosis. The presence of two antibody classes with reactivity toward the basement membrane zone may have implications for disease severity and treatment considerations. Positive IgA basement membrane zone antibodies may be useful markers for following disease expression and activity, and, based on the presence of IgA epithelial antibodies, dapsone therapy may be indicated (if glucose-6-phosphate dehydrogenase, G6PD, enzymatic activity in blood is normal).

TESTING METHODS

Indirect Immunofluorescence (IIF)

IgA Epithelial Basement Membrane Zone (BMZ) Antibodies

Result Footnote

f1: Epithelial BMZ Ab, IgA
Patient serum is progressively diluted beginning at 1:5 in three two-fold screening dilutions, layered on sections of human skin split at the basement membrane zone and monkey esophagus substrates, and reacted with fluorescein isothiocyanate (FITC)-conjugated antibody to IgA. When positive, the serum is further diluted in two-fold reductions to the limiting dilution of antibody detection or to a maximum dilution of 1:40,960. The limiting-dilution, end-point titer is reported for each substrate, and the pattern of staining on split skin substrate also is reported. This indirect immunofluorescence testing was developed and its performance characteristics determined by the Immunodermatology Laboratory at the University of Utah. It has not been cleared or approved by the FDA (US Food and Drug Administration). FDA clearance or approval currently is not required for this testing performed in a CLIA-certified laboratory (Clinical Laboratory Improvement Amendments) and intended for clinical use. [Indirect immunofluorescence, one antibody on two substrates (IIF X 2) with two limiting-dilution, end-point titers (antibody titer X 2)]

Electronically signed by Kristin M. Leiferman, MD, on 06/22/22 at 1:42 PM.
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