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PATIENT REPORT

Patient Age/Sex: 28 years Female

Specimen Collected: 22-Jun-22 13:21

Pemphigus Antibody Panel, IgG	**Received:**		**Report/Verified: 22-Jun-22 13:55**
Procedure	**Result**	**Units**	**Reference Interval**
EER Pemphigus Antibody Panel,IgG	See Note [f1]		

Pemphigus Antibody Panel, IgG	**Received: 22-Jun-22 13:59**		**Report/Verified: 22-Jun-22 15:39**
Procedure	**Result**	**Units**	**Reference Interval**
Pemphigus Antibody Panel,IgG	See Note [f2]		

Result Footnote

f1: EER Pemphigus Antibody Panel, IgG
Authorized individuals can access the ARUP Enhanced Report using the following link:

f2: Pemphigus Antibody Panel, IgG
CLINICAL INFORMATION
Mucosal erosions and scattered eroded lesions on upper body. Presumptive diagnosis is pemphigus versus pemphigoid.

Specimen Details
S22-IP0000514 - Serum; Collected: 6/22/2022; Received: 6/22/2022

DIAGNOSTIC INTERPRETATION

Consistent with pemphigus vulgaris

(See Results and Comments)

RESULTS
Indirect Immunofluorescence (IIF)

Cell Surface (CS)/Intercellular Substance (ICS) IgG Antibodies

IgG: Positive, titer 1:5,120 (H), monkey esophagus substrate
Positive, titer 1:2,560 (H), intact human skin substrate

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

(H) = high/positive

Enzyme-Linked Immunosorbent Assay (ELISA)

Desmoglein (DSG) 1 and 3 IgG Antibodies

IgG desmoglein 1 antibody level: 3 U/mL

Reference Range:
Normal (negative) = Less than 14 U/mL

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

Result Footnote

f2: Pemphigus Antibody Panel, IgG
Borderline/Indeterminate = 14-20 U/mL
Increased (H) (positive) = Greater than 20 U/mL

IgG desmoglein 3 antibody level: 98 U/mL (H)

Reference Range:
Normal (negative) = Less than 9 U/mL
Borderline/Indeterminate = 9-20 U/mL
Increased (H) (positive) = Greater than 20 U/mL

COMMENTS

Specific

The indirect immunofluorescence results, demonstrating positive IgG cell surface (CS), also known as intercellular substance (ICS), antibodies, support the diagnosis of pemphigus. The ELISA results, demonstrating an increased IgG desmoglein 3 antibody level and a normal IgG desmoglein 1 antibody level, further support the diagnosis of pemphigus vulgaris. IgG CS/ICS antibody titers by indirect immunofluorescence and IgG desmoglein antibody levels by ELISA correlate with disease activity in pemphigus vulgaris and pemphigus foliaceus.

IgG CS/ICS antibodies characteristically are positive by indirect immunofluorescence in IgG pemphigus variants, including pemphigus foliaceus and pemphigus vulgaris, and IgA CS/ICS antibodies characteristically are positive by indirect immunofluorescence in IgA pemphigus, although IgA CS/ICS antibodies may be observed in some pemphigus variants along with positive IgG CS/ICS antibodies. If indicated to further evaluate for IgA CS/ICS antibodies, add-on testing may be requested on this specimen by contacting ARUP Client Services at 1-800-242-2787, option 2, for:
- Pemphigus Antibodies, IgA by IIF (ARUP test number 0092106).

Clinical correlation is needed, including treatment status, with consideration for monitoring antibody profiles by indirect immunofluorescence and antibody levels by ELISAs to aid in assessing disease expression and activity, including response to therapy.

General

Greater than 80 percent of patients with pemphigus have positive epithelial cell surface (CS), also known as intercellular substance (ICS), antibodies in their sera identified by indirect immunofluorescence. Serum antibody titers correlate with disease activity, and CS/ICS antibodies may be in low titer or negative in patients whose disease activity is minimal and/or under therapeutic control. Cell surface antibodies are implicated in the pathophysiology of pemphigus. However, cell surface reactivity may be observed transiently and/or nonspecifically in normal individuals and in patients with infections, drug reactions, and other mucocutaneous disorders, including other immunobullous diseases, generally in low titer. IgA cell surface antibodies, are positive by indirect immunofluorescence in patients with IgA pemphigus and in some pemphigus variants along with positive IgG cell surface antibodies. Approximately 40 percent of patients with nonclassical IgG/IgA pemphigus have an underlying systemic disease when diagnosed, malignancy being the most common.

Pathogenic antibodies in serum from individuals with pemphigus bind to desmogleins, which are calcium-dependent adhesion molecules in cell surface desmosomes; such antibodies are detected by ELISA. Specific reactivity to the type of desmoglein may be helpful in determining pemphigus subtypes; IgG desmoglein 1 autoantibodies predominate in patients with pemphigus foliaceus, and IgG desmoglein 3

Result Footnote

f2: Pemphigus Antibody Panel, IgG
autoantibodies, with or without accompanying desmoglein 1 autoantibodies, predominate in patients with pemphigus vulgaris. Overlapping expression with autoantibodies to both desmogleins 1 and 3 clinically is associated with both mucosal and skin lesions. ELISA testing for IgG desmoglein 1 and IgG desmoglein 3 antibodies is highly sensitive, with greater than 90 percent of pemphigus patients showing increased levels of one or both antibodies. IgG desmoglein antibody levels also correlate with disease activity in pemphigus foliaceus and pemphigus vulgaris; however, patients with CS/ICS antibody-positive pemphigus by indirect immunofluorescence can have normal results on ELISA testing with epithelial CS/ICS antibodies to different desmoglein 1 and/or desmoglein 3 epitopes than displayed in the tested ELISAs or to other adhesion molecules.

TESTING METHODS
Indirect Immunofluorescence (IIF)

IgG Epithelial Cell Surface (CS)/Intercellular Substance (ICS) Antibodies

Patient serum is progressively diluted in calcium-containing buffer beginning at 1:10 in three two-fold screening dilutions, layered on sections of intact normal human skin and monkey esophagus substrates, and reacted with fluorescein isothiocyanate (FITC)-conjugated antibody to IgG. 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. 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)]

Enzyme-Linked Immunosorbent Assay (ELISA)

IgG desmoglein 1 and IgG desmoglein 3 serum antibody levels determined by U.S. Food and Drug Administration (FDA)-approved ELISAs (Mesacup, MBL BION). [Two ELISAs]

Electronically signed by Kristin M. Leiferman, MD, on 06/22/22 at 3:35 PM.
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