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PATIENT REPORT

Patient Age/Sex: 43 years Female

Specimen Collected: 21-Jun-22 15:46

Epithelial Cell Surface Antibody IgG | **Received: 22-Jun-22 09:55** | **Report/Verified: 22-Jun-22 14:32**

Procedure	Result	Units	Reference Interval
Epithelial Cell Surface Ab IgG	See Note [f1]		

Result Footnote

f1: Epithelial Cell Surface Ab IgG
CLINICAL INFORMATION
Mucosal erosions and few scattered eroded lesions on upper body skin. Presumptive diagnosis is pemphigus versus mucous membrane pemphigoid.

Specimen Details
S22-IP0000505 - Serum; Collected: 6/21/2022; Received: 6/22/2022

DIAGNOSTIC INTERPRETATION

Consistent with IgG-variant pemphigus, including pemphigus foliaceus and pemphigus vulgaris

(See Results and Comments including further testing recommendations)

RESULTS
Indirect Immunofluorescence (IIF)

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

IgG: Positive, titer 1:2,560 (H), monkey esophagus substrate
Positive, titer 1:640 (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

COMMENTS

Specific

These indirect immunofluorescence test results, demonstrating positive serum IgG cell surface (CS), also known as intercellular substance (ICS) antibodies, reactive with both monkey esophagus substrate and intact human skin substrate, support the diagnosis of pemphigus vulgaris or pemphigus foliaceus or another IgG-variant pemphigus. Pemphigus foliaceus and pemphigus vulgaris can be distinguished by the predominance of IgG desmoglein 1 or IgG desmoglein 3 antibodies, respectively, determined by enzyme-linked immunosorbent assay (ELISA). Another consideration is that, although IgA CS/ICS antibodies characterize IgA pemphigus, they also may be observed in some pemphigus variants along with positive IgG CS/ICS antibodies.

Further ELISA testing for IgG desmoglein 1 and IgG desmoglein 3 antibody levels is recommended and can be accomplished on this specimen by contacting ARUP Client Services, 1-800-242-2787, option 2, with add-on test request for:

Result Footnote

f1: Epithelial Cell Surface Ab IgG
- Desmoglein 1 and Desmoglein 3 (Pemphigus) Antibodies, IgG by ELISA (ARUP test number 0090649),

with or without additional indirect immunofluorescence testing to determine if IgA CS/ICS antibodies also are expressed with add-on test request for:
- Pemphigus Antibodies, IgA by IIF (ARUP test number 0092106).

IgG cell surface antibody titers and IgG desmoglein antibody levels correlate with disease activity in IgG-variant pemphigus, including pemphigus foliaceus and pemphigus vulgaris. Of note, cell surface reactivity by indirect immunofluorescence may be observed transiently and/or nonspecifically in normal individuals, and in patients with infections, drug reactions, and other mucocutaneous disorders. Clinical correlation is needed, including treatment status and with direct immunofluorescence findings on a biopsy specimen. Monitoring antibody levels by ELISAs as well as antibody profiles by indirect immunofluorescence may 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) antibodies, 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. 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 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. Approximately 40 percent of patients with nonclassical IgG/IgA pemphigus have an underlying systemic disease when diagnosed, malignancy being the most common.

Pathogenic IgG CS/ICS antibodies in serum from patients with pemphigus bind to desmogleins, which are calcium-dependent adhesion molecules in desmosomes; such antibodies are detected by enzyme-linked immunosorbent assay (ELISA) testing. Specific reactivity to the type of desmoglein may be helpful in determining pemphigus subtypes; the IgG desmoglein 1 antibody level is increased in patients with pemphigus foliaceus, and the IgG desmoglein 3 antibody level, with or without an increased IgG desmoglein 1 antibody level, is predominantly increased in patients with pemphigus vulgaris. Overlapping expression with autoantibodies to both desmogleins 1 and 3 typically is associated clinically 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 patients with pemphigus showing increased levels of one or both antibodies.

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 stained 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 IIF testing was developed and its performance characteristics determined by the Immunodermatology Laboratory

Result Footnote

f1: Epithelial Cell Surface Ab IgG
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 2:30 PM.
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