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PATIENT REPORT

Patient Age/Sex: 19 years Female

Specimen Collected: 21-Jun-22 15:48

Paraneoplastic Pemphigus Ab Screen **|Received: 22-Jun-22 09:57** **Report/Verified: 22-Jun-22 15:02**

Procedure	Result	Units	Reference Interval
Paraneoplastic Pemphigus Ab Screen	See Note [f1]		

Result Footnote

f1: Paraneoplastic Pemphigus Ab Screen
CLINICAL INFORMATION
Mucosal involvement with extensive erosions and targetoid, urticarial, and scaly skin lesions.
Presumptive diagnosis is drug reaction versus pemphigus, including paraneoplastic pemphigus.

Specimen Details
S22-IP0000507 - Serum; Collected: 6/21/2022; Received: 6/22/2022

DIAGNOSTIC INTERPRETATION

Positive findings, consistent with paraneoplastic pemphigus

(See Results and Comments including further testing considerations)

RESULTS
Indirect Immunofluorescence (IIF)

Paraneoplastic Pemphigus IgG Antibodies

IgG: Positive, titer 1:10,240 (H), rat bladder substrate
(cell surface)
Positive, titer 1:640 (H), rat bladder substrate
(basement membrane zone)

Positive, titer 1:2,560 (H), mouse bladder substrate
(cell surface)
Positive, titer 1:1,280 (H), mouse bladder substrate
(basement membrane zone)

Positive, titer 1:40 (H), mouse heart substrate
(intercalated discs)
Positive, titer 1:160 (H), mouse liver substrate
(portal tracts)

Reference Range:
Negative - Titer less than 1:5
Borderline - Titer 1:5
Positive (H) - Titer greater than 1:5

Positive, monkey esophagus substrate
(cell surface)
Negative, monkey esophagus substrate
(basement membrane zone)

Reference Range:

Result Footnote

f1: Paraneoplastic Pemphigus Ab Screen
Negative - Titer less than 1:10
Borderline - Titer 1:10
Positive (H) - Titer greater than 1:10

(H) = high/positive

COMMENTS

Specific

IgG antibodies reactive with rodent substrates, including cell surface and basement membrane zone antibodies with rat and mouse bladder substrates and cell surface antibodies with monkey esophagus substrate, as detected in this indirect immunofluorescence testing, are consistent with the diagnosis of paraneoplastic pemphigus, also known as Paraneoplastic Autoimmune Multiorgan Syndrome (PAMS). Antibody reactivity with intercalated discs in rodent heart substrate and portal tracts in rodent liver substrate is supportive when antibody reactivity with rodent bladder substrate, either or both rat and mouse, is present.

Various serum epithelial antibodies may be found in paraneoplastic pemphigus and other paraneoplastic presentations by various tests with differing sensitivities. Moreover, detection, levels, and patterns of diagnostic antibodies may fluctuate with disease manifestations. Correlation with clinical presentation, direct immunofluorescence findings on a biopsy specimen, histopathological examination of formalin-fixed tissue, and other epithelial antibodies in serum is recommended. To further evaluate the immunopathological profile, additional testing may be performed on this serum specimen by contacting ARUP Client Services, 1-800-242-2787, option 2, with add-on test request(s) for:
- Pemphigus Antibody Panel, IgG (ARUP test number 0090650); and/or
- Pemphigus Antibodies, IgA by IIF (ARUP Test number 0092106); and/or
- Basement Membrane Zone Antibody Panel (ARUP test number 3001410).

Or the test panel that includes all of the above:
- Immunobullous Disease Antibody Panel (ARUP test number 3001409).

Monitoring serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs may aid in assessing disease expression and activity, including therapeutic response.

General

Positive Paraneoplastic Pemphigus Antibody Screen testing results by indirect immunofluorescence indicate the presence of serum antibodies to multiple epithelia (simple, columnar, transitional) with several possible epithelial targets, predominantly to plakins (envoplakin, periplakin, desmoplakin I, desmoplakin II, epiplakin, plectin, BP230), also cadherins (desmoglein 1, desmoglein 3; desmocollin 1, desmocollin 2, desmocollin 3), alpha-2-macroglobulin-like-1 (A2ML1), laminin-332, and/or BP180 and support a diagnosis of paraneoplastic pemphigus (paraneoplastic autoimmune multiorgan syndrome). For positive antibody screen testing results without known malignancy, perform aggressive evaluation for malignancy.

Paraneoplastic pemphigus (paraneoplastic autoimmune multiorgan syndrome) may affect all ages and develops as a severe mucocutaneous blistering and erosive disease in association with malignancies, most often hematologic (lymphoma, leukemia) and sarcoma. It also may develop in association with benign neoplasias, especially Castleman disease, which is the most frequent association in children and adolescents. Antibodies targeting the various types of epithelia can lead to involvement of various organs and

Result Footnote

f1: Paraneoplastic Pemphigus Ab Screen
tissues, for example, eyes, lungs, gastrointestinal tract, kidney, and thyroid and is the basis of the name, paraneoplastic autoimmune multiorgan syndrome.

Negative Paraneoplastic Pemphigus (Paraneoplastic Pemphigus Autoimmune Multiorgan Syndrome) Antibody Screening results by indirect immunofluorescence do not rule out paraneoplastic/malignancy-associated disease. For negative results, correlate with findings by histopathological examination of formalin-fixed tissue in addition to direct immunofluorescence testing on a biopsy specimen and serum epithelial antibodies characteristic of other immunobullous diseases with further clinical evaluation as indicated.

TESTING METHODS
IgG Paraneoplastic Pemphigus Antibodies

The patient serum is progressively diluted in calcium-containing buffer beginning at 1:5 in three two-fold screening dilutions, layered on rodent substrates, including rat bladder, mouse bladder, mouse heart, and mouse liver, 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 rodent 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 four rodent substrates and one antibody on monkey esophagus substrate (IIF X 5) with six limiting dilution, end-point titers (antibody titer X 6)]

Electronically signed by Kristin M. Leiferman, MD, on 06/22/22 at 3:00 PM.
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