

Client: Example Client ABC123 123 Test Drive Salt Lake City, UT 84108 UNITED STATES

Physician: Doctor, Example

Patient: Patient, Example

DOB 12/31/1965 Gender: Female

Patient Identifiers: 01234567890ABCD, 012345

Visit Number (FIN): 01234567890ABCD **Collection Date:** 00/00/0000 00:00

Direct Immunofluorescence, Tissue Biopsy (Cutaneous, Mucosal, Epithelial)

ARUP test code 0092572

EER Cutaneous Direct IF, Biopsy

See Note

Authorized individuals can access the ARUP Enhanced Report using the following link:

Cutaneous Direct IF, Biopsy

See Note

CLINICAL INFORMATION Blisters on urticarial base for 3 months with occasional oral lesions, evaluate for pemphigoid and other immunobullous diseases

Specimen Details

Left arm, punch, perilesional, sun-exposed;
Collected: 2/2/2024; Received: 2/6/2024

- Right buccal mucosa, punch, perilesional;
Collected: 2/2/2024; Received: 2/6/2024

DIAGNOSTIC INTERPRETATION

Consistent with pemphigoid and pemphigoid variants, including possible lichen planus pemphigoides

RESULTS

Examination of sun-exposed left arm perilesional skin punch biopsy (Specimen A) and right buccal perilesional oral mucosa punch biopsy (Specimen B) tissue sections tested for immunoglobulins, complement, and fibrinogen reveals:

Specimen A

IgG: 3+ linear basement membrane zone IgG4: 2+ linear basement membrane zone

Specimen B

IgG: 3+ discontinuous linear basement membrane zone IgG4: 3+ linear basement membrane zone

Specimen A

IgM: 2+ few scattered and clumped cytoids Specimen B

IgM: 3+ several scattered and clumped cytoids

Specimen A

H=High, L=Low, *=Abnormal, C=Critical



Scattered inflammatory cells, apparent eosinophils IaA:

Specimen B

Negative IaA:

Specimen A
C3: 3+ linear basement membrane zone and scattered

inflammatory cells, apparent eosinophils

Specimen B

2+ linear to focal granular basement membrane C3:

zone

Specimen A

Fibrinogen: 2+ discontinuous linear basement membrane

zone and 3+ patchy deposition on upper and mid dermal connective tissue fibers

Specimen B

Fibrinogen: 2-3+ linear to shaggy basement membrane

zone, 3+ few scattered and clumped cytoids, and 3+ deposition on

subepithelial connective tissue fibers

These direct immunofluorescence findings are consistent with pemphigoid and pemphigoid variants, including epidermolysis bullosa acquisita, based on linear IgG, including IgG4, basement membrane zone antibody reactivity and linear C3 basement membrane zone localization. Apparent eosinophil infiltration also is present in the skin biopsy tissue (Specimen A) as often observed in pemphigoid. Shaggy fibrinogen basement membrane zone deposition and cytoid bodies in the mucosal biopsy tissue (Specimen B) further are features of a lichenoid reaction as found in lichen planus pemphigoides.

Pemphigoid, epidermolysis bullosa acquisita, and other pemphigoid variants cannot be distinguished by direct immunofluorescence; however, if circulating antibodies are present, these diseases can be distinguished by indirect immunofluorescence based on the localization pattern of serum basement membrane zone antibodies with human split skin substrate (also known as salt split skin). In addition, IgG BP180 and IgG BP230 antibody levels, as determined by enzyme-linked immunosorbent assays (ELISA), are diagnostic markers for pemphigoid, and the IgG type VII collagen antibody level, as determined by ELISA, is a diagnostic marker for epidermolysis bullosa acquisita. Serum basement membrane zone antibodies are detected in up to 80 percent of patients with these disorders, and antibody levels by ELISAS may be useful in monitoring disease activity and response to therapy. monitoring disease activity and response to therapy.

Correlation with clinical presentation is needed. To further define the diagnostic immunopathological expression profile and for monitoring disease activity, correlation with serum basement membrane zone antibody testing is recommended and may be accomplished by submitting a serum specimen through ARUP Laboratories with request for the Basement Membrane Zone Antibody Panel (ARUP test number 3001410); contact ARUP Client Services, 1-800-242-2787, option 2, for assistance, if needed.

Lichenoid reactions characteristically are found in lichen planus, and also may be found in association with epithelial malignancies or premalignancy, and in lupus erythematosus, drug reactions, erythema multiforme, and other mucocutaneous disorders including lichen planus pemphigoides and paraneoplastic pemphigus. Direct immunofluorescence is not useful for diagnosis of malignancies involving the skin or mucous membranes. Lichenoid reactions in mucosa most commonly indicate lichen planus, and, in this case, lichen planus pemphigoides. Correlation with histopathological examination of formalin-fixed tissue is needed.

High resolution, color digital images of representative direct

H=High, L=Low, *=Abnormal, C=Critical

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immunofluorescence findings are available for this testing (see images in the Enhanced Electronic Report/EELR). If you would like a hard copy or an electronic file of the images and/or if it would be helpful to discuss the patient case with this report, contact ARUP Client Services at 1-800-242-2787, option 2, and ask to speak with the Immunodermatology Laboratory at the University of Utah regarding patient results.

TESTING METHODS

The tissue specimens from perilesional left arm skin (Specimen A) and right buccal oral mucosa (Specimen B) received in Michel transport medium, are each washed, cryoembedded into blocks, cryosectioned, and reacted with fluorescein isothiocyanate (FITC)-conjugated antibodies to IgG, IgG4, IgM, IgA, C3, and fibrinogen. IgG4 subclass staining is performed because IgG4 reactivity may be more sensitive than IgG in some immune-mediated diseases. The stained tissue sections are examined by fluorescence microscopy to identify patterns of positive reactivity that aid in the diagnosis of immune-mediated diseases. Positive findings are graded on a visual analog scale of 0-3+ (weak - detected; 1+ - definite, but relatively slightly, positive; 2+ - moderately positive intensity; 3+ - strongly positive). Negative control serial sections exposed to bovine serum albumin (BSA) without antibody and a technically adequate hematoxylin and eosin (H and E)-stained slide are prepared and also examined for comparison to specific staining and for morphological orientation and features. This direct immunofluorescence (DIF) 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.

Electronically signed by ■ 7:08 PM.

on 02/11/24 at

7.08 PM.
Performed At: IMMUNODERMATOLOGY LABORATORY
417 S. WAKARA WAY, SUITE 2151
SALT LAKE CITY, UT 84108
Medical Director: KRISTIN M. LEIFERMAN, MD
CLIA Number: 46D0681916

VERIFIED/REPORTED DATES				
Procedure	Accession	Collected	Received	Verified/Reported
EER Cutaneous Direct IF, Biopsy	24-033-117722	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00
Cutaneous Direct IF, Biopsy	24-033-117722	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00

END OF CHART

H=High, L=Low, *=Abnormal, C=Critical

Patient: Patient, Example ARUP Accession: 24-033-117722 Patient Identifiers: 01234567890ABCD, 012345 Visit Number (FIN): 01234567890ABCD Page 3 of 3 | Printed: 10/29/2024 9:17:52 AM 4848