Test Definition: PNPAB

Paraneoplastic Pemphigus Antibody, IgG, Serum

Overview

Useful For

Diagnosis of paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome in the setting of erosive or lichenoid mucocutaneous disease

Testing Algorithm

For information see Autoimmune Blistering Disease Diagnostic Algorithm.

Special Instructions

- Autoimmune Blistering Disease Diagnostic Algorithm

Method Name

Indirect Immunofluorescence Assay (IFA)

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

Supplies: Sarstedt Aliquot Tube, 5 mL (T914)
Collection Container/Tube:
Preferred: Serum gel
Acceptable: Red top
Submission Container/Tube: Plastic vial
Specimen Volume: 2 mL Serum
Collection Instructions: Centrifuge and aliquot serum into a plastic vial.

Specimen Minimum Volume

Serum: 0.5 mL

Reject Due To

Gross hemolysis	Reject
Gross lipemia	Reject

Gross icterus	Reject

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	14 days	
	Ambient	14 days	
	Frozen	30 days	

Clinical & Interpretive

Clinical Information

Paraneoplastic pemphigus (PNP) (also paraneoplastic autoimmune multiorgan syndrome [PAMS] to denote the systemic nature of the syndrome) is an autoimmune mucocutaneous blistering disease affecting adults or, rarely, children that generally heralds the presence of an underlying malignancy.

PNP/PAMS can be defined and identified by a combination of the following features:
1. Painful stomatitis and a polymorphous cutaneous eruption with lesions that may be blistering, lichenoid, erythema multiforme-like, or morbilliform
2. Variable histopathologic findings, including acantholysis, lichenoid, or interface change
3. Variable direct immunofluorescence findings from a perilesional biopsy, often demonstrating deposition of IgG and complement in the epidermal intercellular spaces, granular/linear complement deposition along the epidermal basement membrane zone, or a lichenoid tissue reaction
4. Indirect immunofluorescence evidence of cell surface deposition on primate esophagus or rat bladder epithelium
5. Enzyme-linked immunosorbent assay evidence of serum autoantibodies against desmogleins 1 or 3 and, possibly, against bullous pemphigoid 180 and 230 antigens
The incidence of the disease is unknown, but it is less common than pemphigus vulgaris or foliaceus. Clinical features of the disease can mimic those seen in a drug reaction, erythema multiforme, Stevens-Johnson syndrome, pemphigus, lichen planus, or toxic epidermal necrolysis.

In the majority of cases, PNP/PAMS is associated with non-Hodgkin lymphoma, chronic lymphocytic leukemia, thymoma, or Castleman disease. A serious complication includes bronchiolitis obliterans, which may lead to respiratory failure.

Reference Values

Negative

Interpretation

In the appropriate clinical setting, a positive result can support a diagnosis of paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome (PNP/PAMS). However, correlation with clinical features, histopathologic findings, results of serum studies (such as indirect immunofluorescence on primate esophagus substrate and enzyme-linked immunosorbent assay for Dsg1/3) is required for a final diagnosis.

As the test is not entirely sensitive, a negative test result does not exclude the possibility of PNP/PAMS.

Cautions

Test results must be interpreted in the patient's individual clinical context.

Clinical Reference

1. Anhalt GJ, Kim SC, Stanley JR, et al. Paraneoplastic pemphigus. An autoimmune mucocutaneous disease associated with neoplasia. N Engl J Med. 1990;323(25):1729-1735. doi:10.1056/NEJM199012203232503
2. Anhalt GJ, Aris-Abdo L, Bonitz P, Labib RS. Antigen specificity of paraneoplastic pemphigus: predictive value of diagnostic techniques based on the study of 17 patients and 135 control subjects. J Invest Dermatol. 1992(4);98:580. Abstract 172
3. Liu AY, Valenzuela R, Helm TN, Camisa C, Melton AL, Bergfeld WF. Indirect immunofluorescence on rat bladder transitional epithelium: a test with high specificity for paraneoplastic pemphigus. J Am Acad Dermatol. 1993;28(5 Pt 1):696-699. doi:10.1016/0190-9622(93)70095-b
4. Camisa C, Helm TN. Paraneoplastic pemphigus is a distinct neoplasia-induced autoimmune disease. Arch Dermatol. 1993;129(7):883-886
5. Montagnon CM, Tolkachjov SN, Murrell DF, Camilleri MJ, Lehman JS. Intraepithelial autoimmune blistering dermatoses: Clinical features and diagnosis. J Am Acad Dermatol. 2021;84(6): 1507-1519. doi:10.1016/j.jaad.2020.11.075
6. Montagnon CM, Lehman JS, Murrell DF, Camilleri MJ, Tolkachjov SN. Intraepithelial autoimmune bullous dermatoses disease activity assessment and therapy. J Am Acad Dermatol. 2021;84(6):1523-1537. doi:10.1016/j.jaad.2021.02.073

Performance

Method Description

Commercially-prepared sections of rat bladder (substrate) are overlaid with patient’s serum; incubated, covered with fluorescein-conjugated IgG antiserum, and interpreted using fluorescent microscopy.(Unpublished Mayo method)

PDF Report

No

Day(s) Performed

Wednesday through Friday

Report Available

7 days

Specimen Retention Time

30 days

Performing Laboratory Location

Mayo Clinic Laboratories - Rochester Main Campus

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Paraneoplastic Pemphigus Antibody, IgG, Serum

Fees & Codes

Fees

- Authorized users can sign in to Test Prices for detailed fee information.
- Clients without access to Test Prices can contact Customer Service 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact Customer Service.

Test Classification

This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. It has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information

86255

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
PNPAB	Paraneoplastic Pemphigus, IgG Ab, S	93233-5

Result ID	Test Result Name	Result LOINC® Value
61881	Paraneoplastic Pemphigus, IgG Ab, S	93233-5