

Plasminogen Deficiency, Type 1: For Ophthalmology



Overview of Plasminogen Deficiency (PLGD)

- PLGD Type 1: a quantitative protein deficiency, with decreased plasminogen activity and decreased antigen
 - Due to a genetic mutation in the gene for plasminogen; > 50 different mutations have been identified; Autosomal Recessive inheritance
 - Most common presenting symptom is ligneous conjunctivitis, but multi-organ, systemic disease that can be life-threatening
- PLGD Type 2: reduced functional activity of plasminogen, but normal antigen levels; patients are asymptomatic

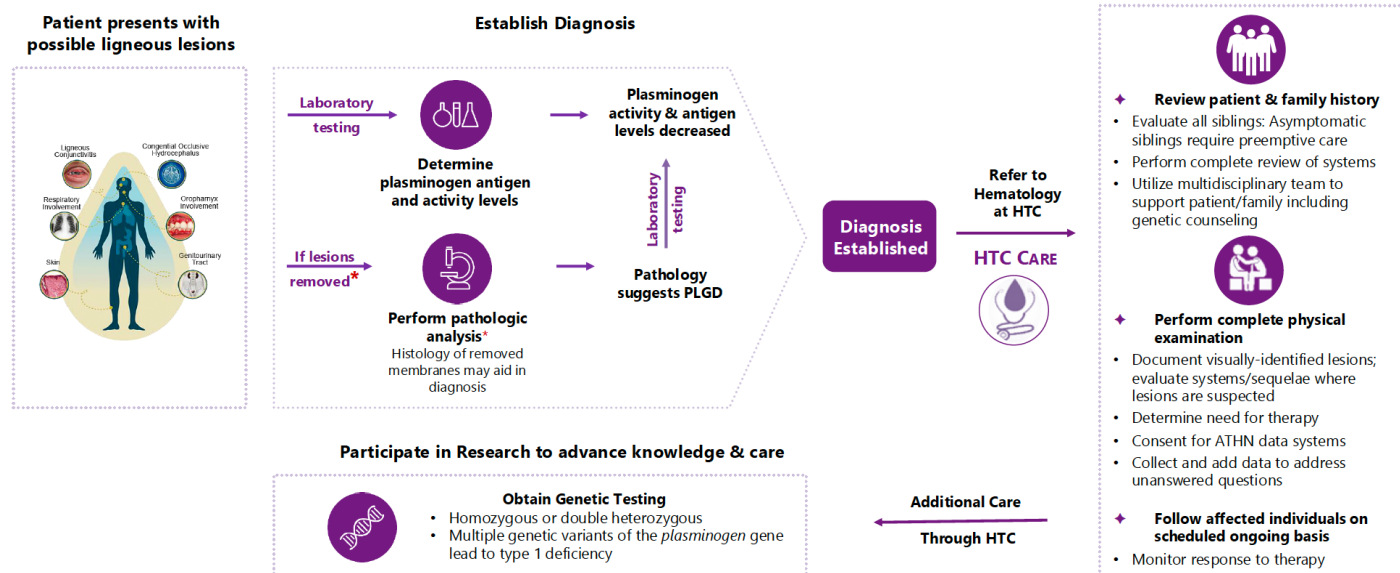
	Normal	PLGD Type 1	PLGD Type 2
Plasminogen Activity	70-130%	Decreased	Decreased
Plasminogen Antigen	6-25 mg/dL	Decreased	Normal
For Patients: My Plasminogen Activity			

Diagnosis

- Complicated by heterogeneous symptoms; symptoms can wax and wane
- Mucosal surfaces of the eyes, ears, nose, gums, airways, lungs, GI tract, kidneys, GU tract, CNS, and skin can all be affected
- Initial point of medical contact therefore includes many disciplines

Treatment

- Ryplazim (plasminogen, human-tvmh) given by IV infusion leads to resolution of lesions
- Surgical removal of lesions, though initially helpful, leads to accelerated regrowth
- Referral to a Hemophilia Treatment Center (HTC) to serve as medical home, and:
 - Educate on product use
 - Ongoing symptom monitoring, outcomes, safety
 - Administer doses, determine dosing schedule
 - Teach home infusion



*Lesion removal prior to diagnosis not recommended

Ophthalmology Specific Diagnostic Findings

INITIAL PRESENTATION		ADVANCED DISEASE
◆ Purulent eye discharge ◆ Excessive tearing ◆ Erythema ◆ Photophobia ◆ Conjunctival inflammation ◆ Ligneous conjunctivitis • Recurrent • Chronic	Untreated 	◆ Chronic fibrotic lesions ◆ Pain ◆ Corneal abrasion & scarring ◆ Visual impairment & amblyopia ◆ Blindness
 Ligneous conjunctivitis		 Advanced ligneous conjunctivitis

Schuster et al. 2003, Ref 1. Used with permission

Ophthalmology Specific Treatment Considerations

- Ligneous conjunctivitis is the most common manifestation of PLGD
 - However, PLGD is a multi-organ, systemic disease
 - May be life threatening
- New or suspected diagnosis:
 - Obtain diagnostic blood test (plasminogen activity level) or refer to hematologist to order
 - If confirmed, refer to HTC to establish care and perform thorough review of systems
 - Patients may have more than one system that is affected at presentation or occurring over time
- Confirmed diagnosis:
 - Coordinate clinical care and collaborate closely with existing care team at HTC
 - Send clinical notes and photos
- Removal of lesions without adequate plasminogen treatment may result in
 - Rapid return of lesions
 - Risk of corneal abrasions, visual impairment, amblyopia, vision loss
- Be suspicious of common diagnoses, for example
 - Ligneous conjunctivitis may present as persistent conjunctivitis before typical lesions appear

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