This child had a history of chronic ligneous conjunctivitis with multiple surgical interventions followed by pseudomembrane regrowth. Hydrocephalus was initially identified at 6 months of age and was thought to be nonprogressive by 3 years of age. Hospital admission at 10 years of age revealed serious systemic manifestations. The patient was the third child of first degree consanguineous parents.

**Initial diagnostic**
- Hydrocephalus

**Diagnosis**
- Nonprogressive hydrocephalus, shunt operation not necessary

**Surgical interventions**
- Multiple excisions of the conjunctival pseudomembranous lesions followed by re-growth

**Physical examination**
- Corneal opacity and leukocoria on the right eye, ligneous conjunctivitis of the left eye, ptosis, blepharitis, and ectropion on the left eye, ligneous gingivitis, macrocephaly (head circumference greater than 97th percentile), frontal bossing

**Neurological examination**
- Conscious and cooperative, full range of eye movement

**Lab test results**
- Prothrombin time: 14 sec, activated partial thromboplastin time: 26.2 sec, INR: 1.08, prothrombin time activity: 89%

**Plasminogen level lab results**
- Patient: 41.5%
- Affected sister (gingival hypertrophy and eye infections without any complications): 41%
- Mother: 147.6%
- Father: 117.4%

**Cranial magnetic resonance imaging**
- Cerebellar and vermian atrophy, dilatation of the third and lateral ventricles, diffuse cerebral parenchymal atrophy

**Development and psychometric tests (Stanford-Binet)**
- Mild degree mental retardation

**Electromyography examination**
- Axonal lesion of peroneal branch of the left sciatic nerve with a few regeneration signs

**Genetic testing**
- No PLG gene mutations but 3 homozygous polymorphisms were identified

**Diagnostic inference**
- Type I Plasminogen Deficiency (PLGD)

**Bronchoscopy examination**
- Mucosal irregularities of pharynx, trachea, larynx, and two main bronchi

**Normal levels of plasminogen activity: 70-130%**

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**References:**


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**KEY TAKEAWAYS**

- Hydrocephalus is a potentially life-threatening complication of Plasminogen Deficiency (PLGD).
- PLGD is an ultra-rare genetic disease that can have devastating effects on multiple organ systems.
- Authors recommend to test for PLGD in patients with hydrocephalus as obstruction of the ventriculoperitoneal shunts is possible when such condition is overlooked.