LARYNGEAL OBSTRUCTION IN A CHILD WITH HISTORY OF LIGNEOUS CONJUNCTIVITIS

Literature-Based Case Study: 5-year-old male child

Diagnosed with Type I Plasminogen Deficiency (PLGD) at 3 months of age, this patient experienced ligneous conjunctivitis and chronic lung disease characterized by hypoxemia and recurrent pneumonia. Hypertonic saline and chest physiotherapy provided relief from respiratory symptoms. At 5 years of age, patient was referred to the otolaryngology/head and neck surgery department because of laryngeal lesions, then admitted with life-threatening airway obstruction requiring surgical intervention.

AGE

3 months

Clinical manifestations
Ligneous conjunctivitis, chronic hypoxemia, recurrent pneumonia, and atelectasis

1st Bronchoscopy
Small polypoid masses in the right bronchus

Clinical manifestations
Failure to thrive, positive celiac serology

Upper gastrointestinal endoscopy
Large lesions in the upper airway region almost occluding the airway, large polypoid lesions in the pyloric outlet and in the esophagus

2nd Bronchoscopy
Supraglottic polypoid mass and several polypoid masses in the distal bronchi

Diagnostic
Type I Plasminogen Deficiency (PLGD) and chronic lung disease

Treatment
Hypertonic saline (3%) and chest physiotherapy
The patient started to thrive and had normal $O_2$ saturation levels in room air, but still suffered from chronic hoarseness

Computer tomography scan
Supraglottic narrowing with no involvement of the trachea or major bronchi

Subsequent inpatient treatment (pediatric intensive care unit)
High dose intravenous steroids and steroid inhalation, intravenous FFP (fresh frozen plasma)

Subsequent procedures
Tracheotomy and laser excision of the supraglottic lesions

Flexible bronchoscopy (through the tracheotomy)
Multiple ligneous lesions in distal small airways

Flexible bronchoscopy (FOL)
Supraglottic polypoid masses occupying the right aryepiglottic fold, the arytenoids, the posterior cricoid area, and the left sinus, pyriformis, obstructing the supraglottic area. The vocal folds could not be visualized.

Pathologic examination
Granulation tissue with areas of ulceration with multiple fibrin deposits
No pathogens were found by use of gram stain, ZN, PAS + D, and/or SILVER. FOL indicated regrowth of the ligneous lesion only 1 week following the tracheotomy procedure

4 years

Subsequent clinical manifestations
Hoarseness and dyspnea (respiratory rate-30/min), mild inspiratory stridor, normal oxygen saturation levels

References:

KEY TAKEAWAYS

- Laryngeal obstruction is a potentially life-threatening complication of PLGD which may result in distortion of the airway anatomy and function.
- PLGD is an ultra-rare genetic disease that can have devastating effects on multiple organ systems.
- When the ligneous lesions develop in the supraglottic area, the vocal folds, or the tracheobronchial tree, dysphonia stridor, and potentially life-threatening airway obstruction and pulmonary disease may occur. Because the clinical course may be complicated by viscous fibrin-rich secretions and recurrent laryngeal, tracheal, and bronchial obstructions by ligneous masses, the prognosis in these cases is poor.

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