Novel treatment for a rare lesion of the trachea

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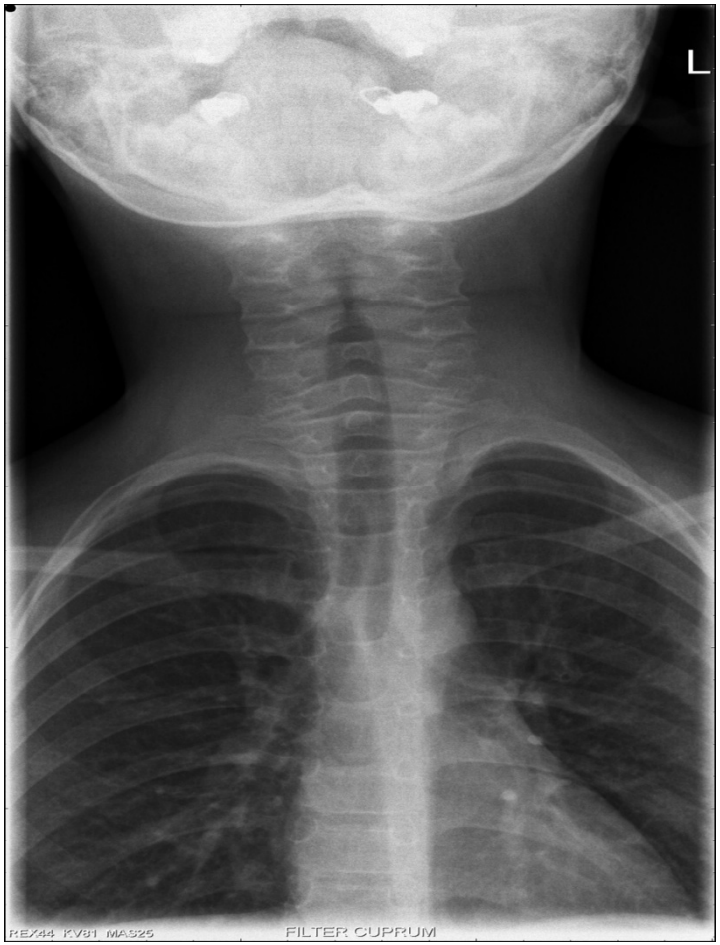
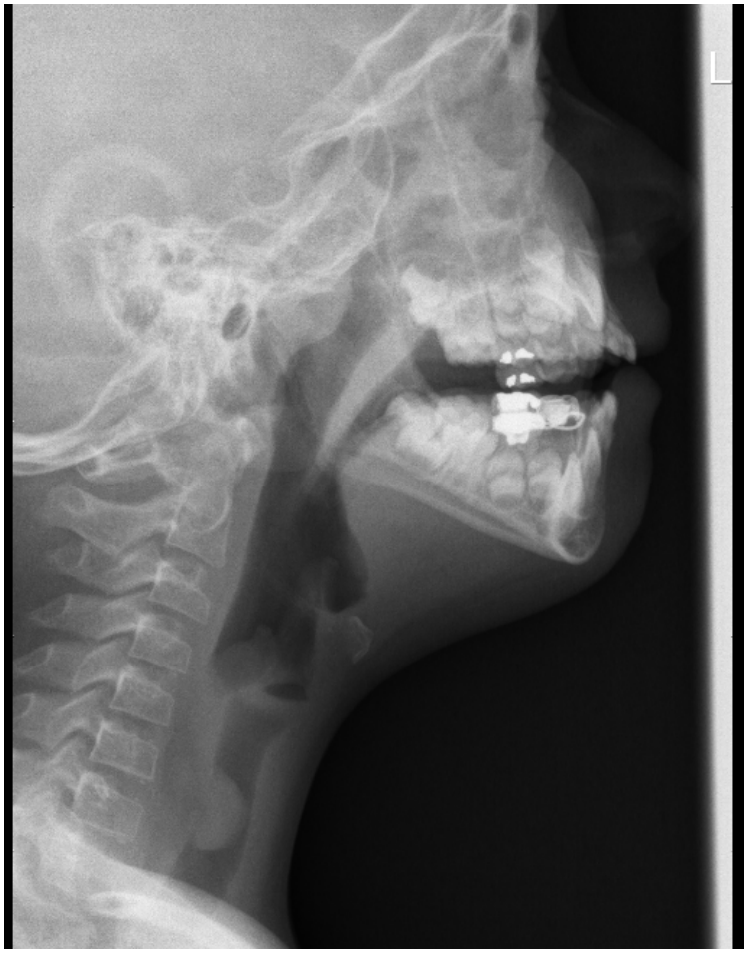
מרכז שניידר לרפואת ילדים בישראל
مركز شنايدر لطب الأطفال في اسرائيل
Schneider Children's Medical Center of Israel

Case Presentation

- 6.7 y/o girl
- No past medical history
- 3 months of respiratory symptoms
- Dyspnea, Stridor, Sleep apnea
- Partial response to bronchodilators and systemic corticosteroids

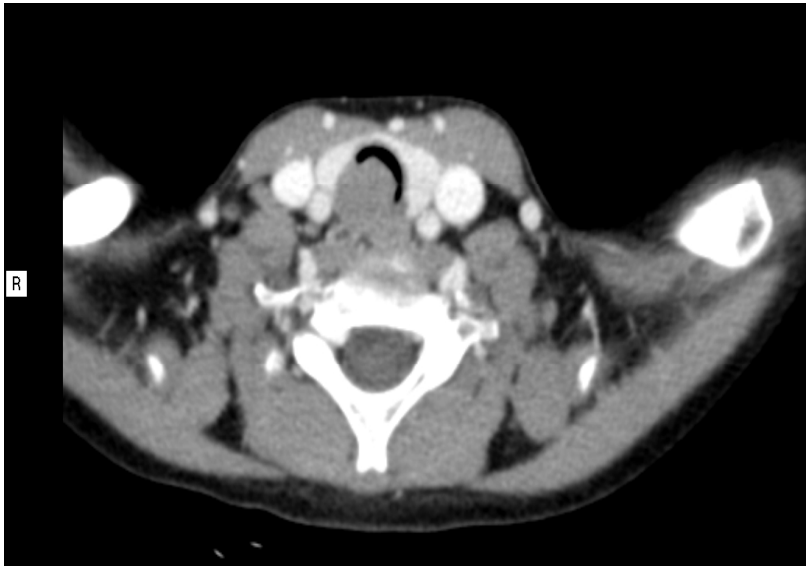
Case Presentation

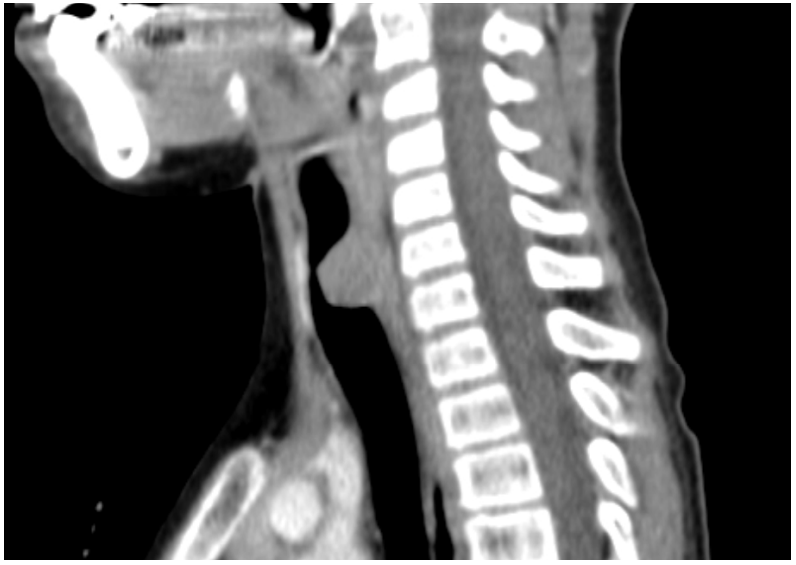




Case Presentation

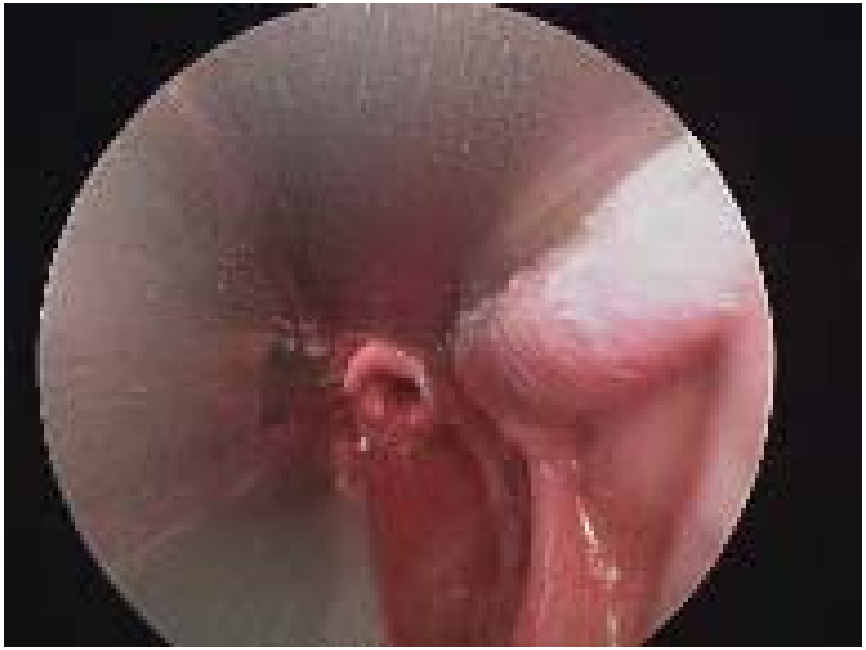
- CT – Mass in Rt posterolateral Tracheal wall



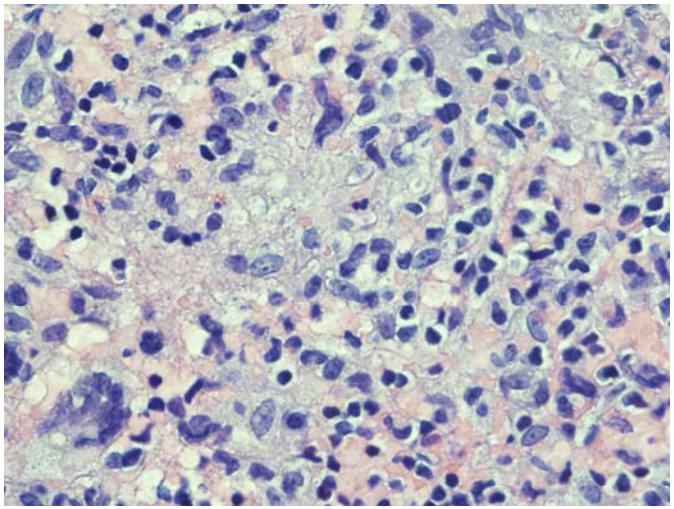


Case Presentation

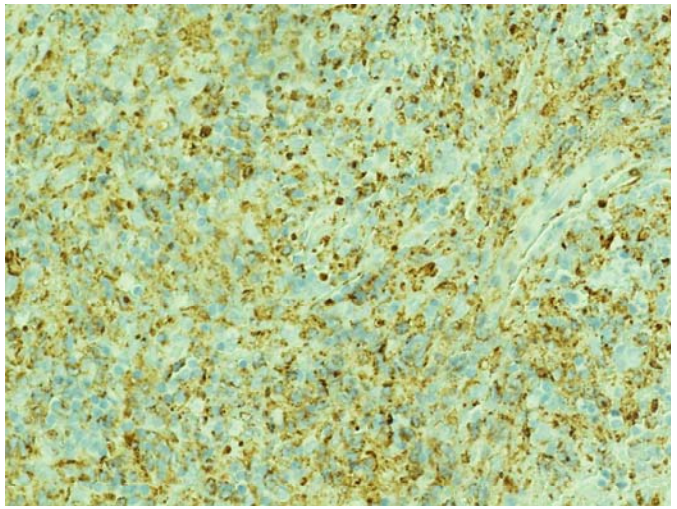
- Bronchoscopy + Bx + Tracheostomy



Pathology



H&E



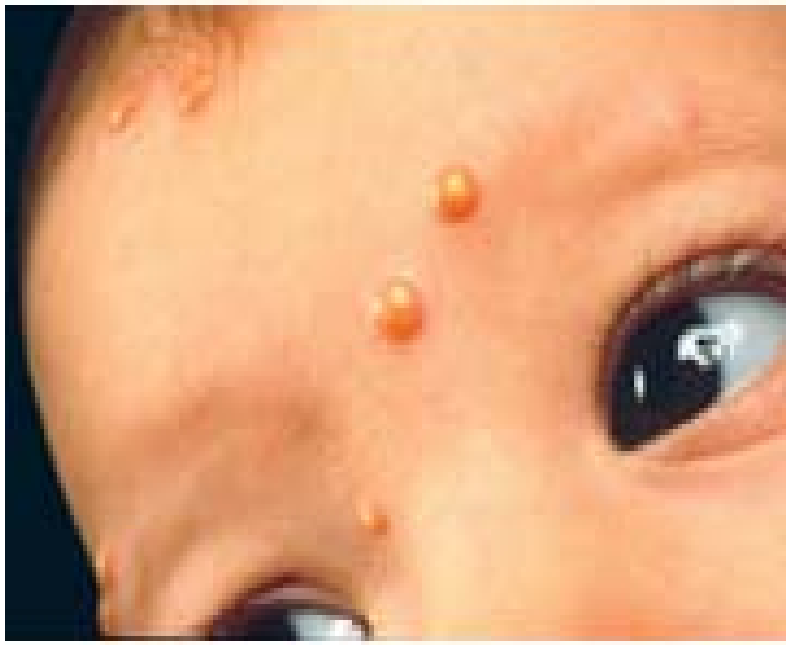
CD-68 Histiocytic marker

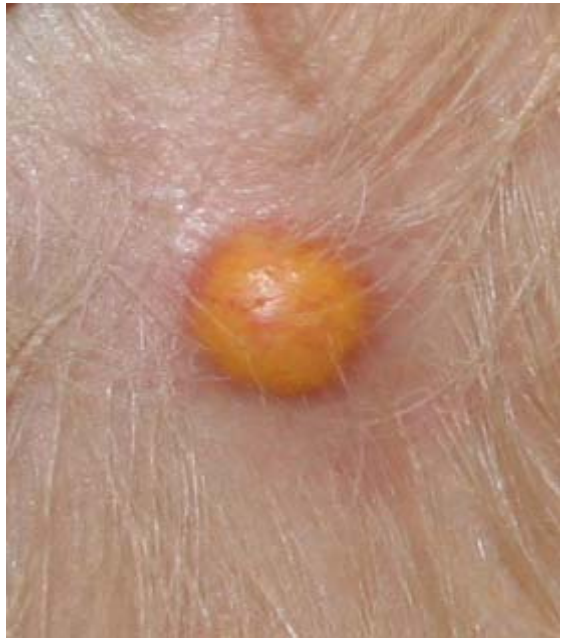
- **Juvenile Xanthogranuloma**

Juvenile Xanthogranuloma

- Self-limited dermatologic disorder
- Associated rarely with systemic manifestations
- Infants and small children are mainly affected
- Lesions may be single or multiple
- Appear as firm, slightly raised papulonodules several millimeters in diameter
- Tan-orange in color
- Occur frequently on the head and neck
- Many extracutaneous sites have been reported

Juvenile Xanthogranuloma





Juvenile Xanthogranuloma

- The etiology is unknown
- Disordered macrophage response to a nonspecific tissue injury
- Granulomatous reaction
- Spectrum of histiocytic disorders
- Less common than Langerhans cell histiocytosis

Juvenile Xanthogranuloma

- Extracutaneous involvement occurs in 4% of children
- Extracutaneous involvement has been reported in every organ system in the body
- CNS, eye, salivary glands, larynx, trachea, lung, pericardium, myocardium, liver, spleen, colon, retroperitoneum, kidney, adrenal gland, gonads, bone, periosteum, muscle, and mucous membranes

JXG Treatment

- Watchful waiting
- Corticosteroids
- Surgery
- Radiotherapy

HEAD & NECK May 2001

ISOLATED JUVENILE XANTHOGRANULOMA OF THE SUBGLOTTIS: CASE REPORT

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Abstract: *Background.* Juvenile xanthogranulomatosis (JXG) is a relatively rare macrophage proliferative disorder. It usually presents as a localized cutaneous lesion but may affect other organs. Until now it has never been described in the subglottic region of the larynx.

Methods. We report the first case of juvenile xanthogranulomatosis (JXG) in the subglottis in a 3 year old child.

Results. The localization in the subglottis caused airway obstruction requiring tracheostomy to secure the airway. On the basis that most cutaneous lesions regress spontaneously the lesion was managed expectantly and regressed over a period of 28 months allowing decannulation of the child.

Conclusion. JXG should be considered in the differential diagnosis of subglottic lesions. Once the airway has been secured, JXG of the subglottis can be managed conservatively. Long-term follow-up is required because of the possibility of relapse at other sites. © 2001 John Wiley & Sons, Inc. *Head Neck* **23:** 426–429 2001.

Keywords: subglottis; juvenile xanthogranulomatosis; larynx, granuloma

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CASE REPORT

Tracheal juvenile xanthogranuloma in a child

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The patient was subsequently managed by external surgical excision. A horizontal suprasternal cervicotomy approach was used. After dissection of the superficial plans an anterior opening of the first 6 tracheal rings was done. The submucosal tumoral mass was excised from second to fourth tracheal ring along with posterior mucous tracheal wall. This wall left open at the end of the procedure. The child was extubated the second day postoperatively with any respiratory impairment.

Pediatric Pulmonology 42:181–185 (2007)

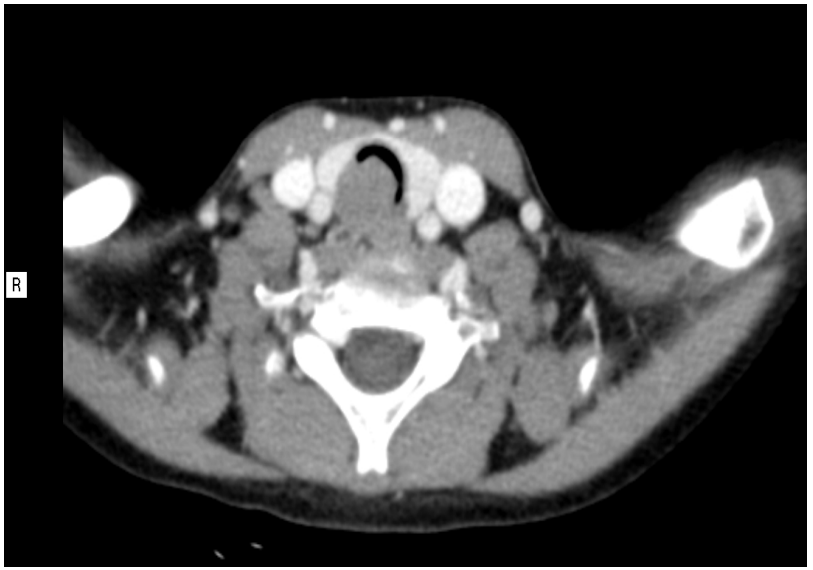
Managing Isolated Subglottic Juvenile Xanthogranuloma Without Tracheostomy: Case Report and Review of Literature

Marta Somorai, MD,[1] Nira A. Goldstein, MD,[2] Richard Alexis, MD,[3] and Robert J. Giusti, MD[1]*

Summary. Juvenile Xanthogranuloma (JXG) is a dendritic cell related histiocytic disorder which usually presents in the first year of life as a solitary cutaneous granuloma. Isolated presentation in the upper airway is very rare but can result in severe respiratory distress, especially in young children. We present the case of a 5-month-old male with an isolated subglottic JXG lesion. Endoscopic excision provided symptomatic relief and avoided the need for tracheostomy. The lesion has completely resolved 17 months later. Surgical excision without tracheostomy was the treatment of choice in two of the four additional cases of upper airway JXG presented in the literature. JXG has an excellent prognosis with spontaneous regression over time. Histology alone is frequently inadequate to differentiate JXG from the more common Langerhans Cell Histiocytosis (LCH), which carries a much less favorable prognosis. The evolving field of immunohistochemistry provides an essential tool to establish the correct diagnosis. The typical phenotype of JXG is Factor XIIIa+/Fascin+/CD68+/CD163+/CD14+/CD1a−/S100−. **Pediatr Pulmonol. 2007; 42:181–185.**

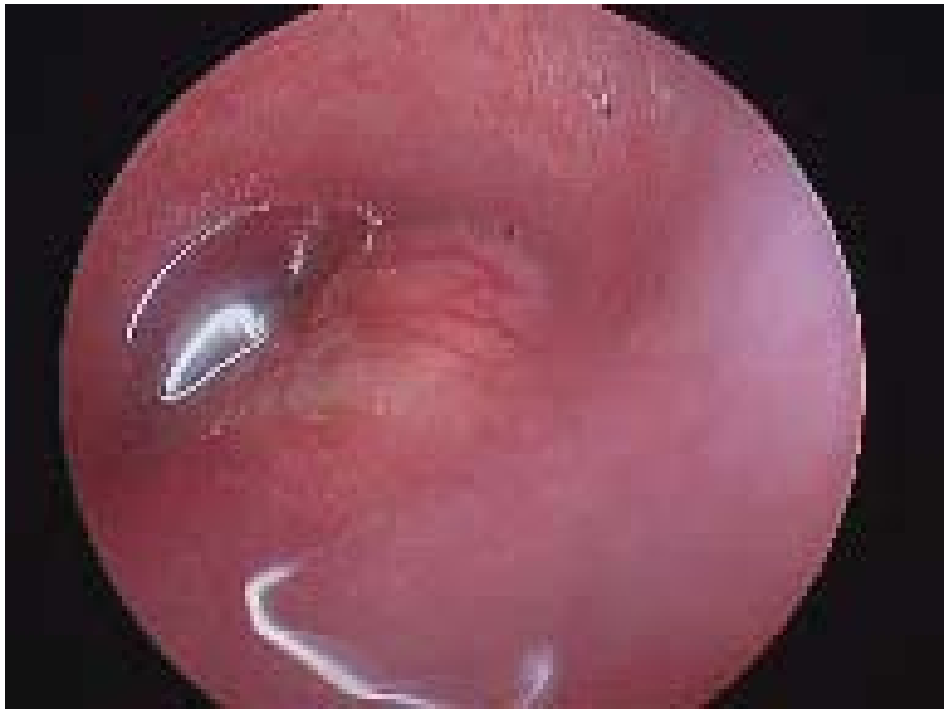
Management

- Watchful waiting
- Surgery
- Radiotherapy
- **Corticosteroids**

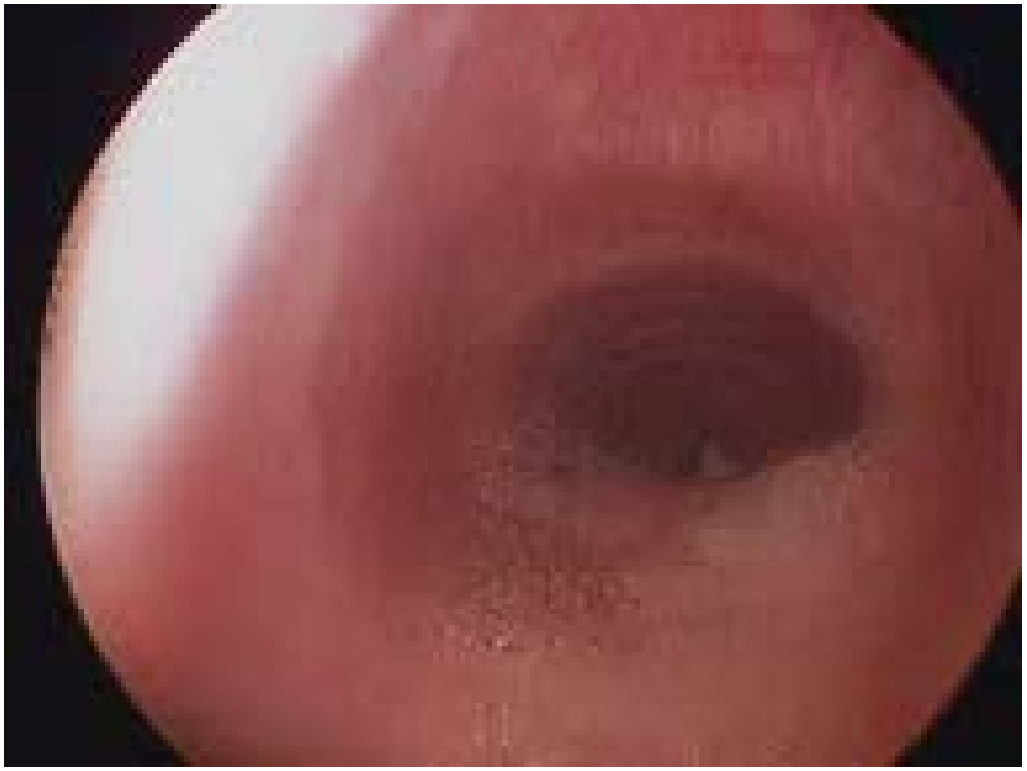


Intralesional Steroid Treatment

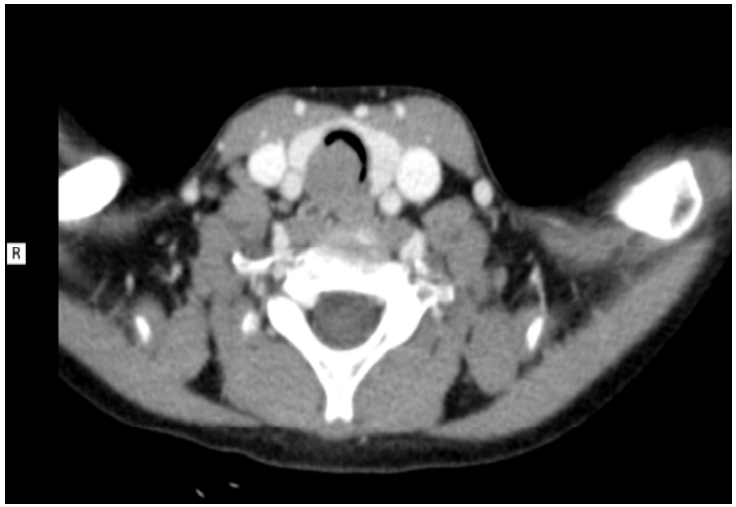
- Methylprednisolone 80mg
- 6 courses

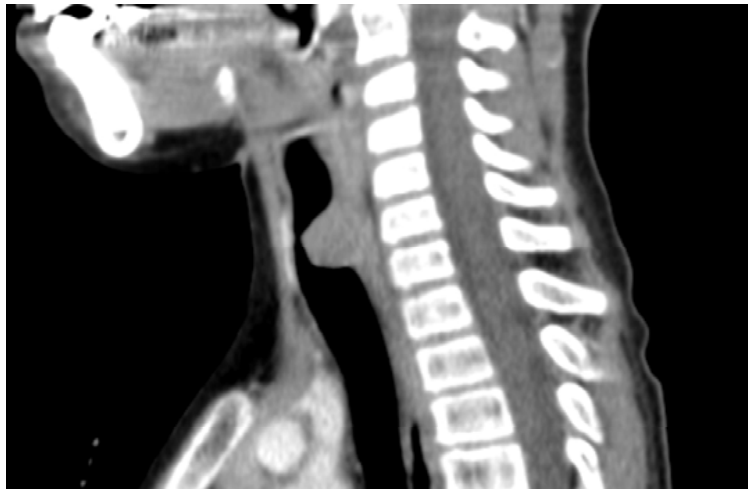


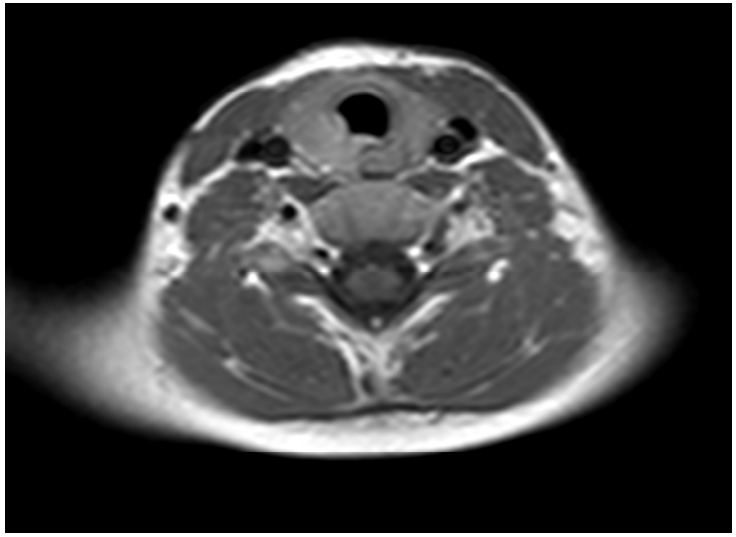
Follow up

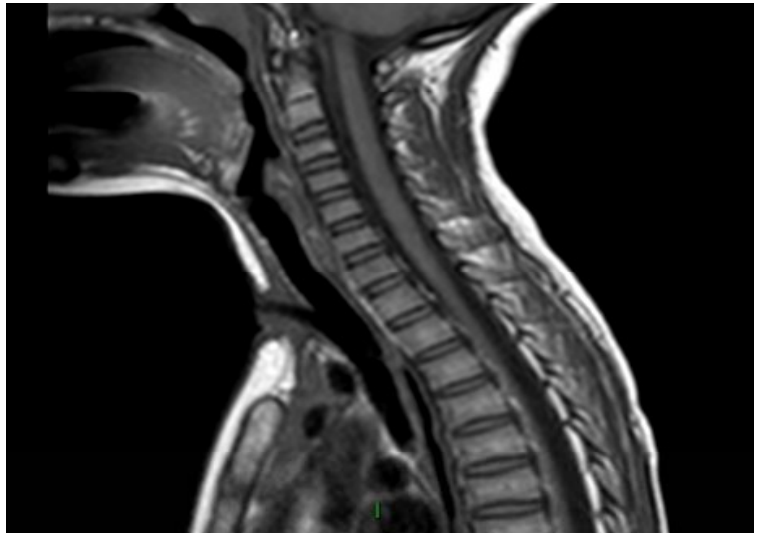


Follow up









Follow up

- Successful Decannulation
- 3 months post treatment
- No respiratory complaints

Thank You