Solitary Juvenile Xanthogranuloma of the Larynx in an Adult: Case Report and Literature Review

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Abstract

Introduction: Juvenile xanthogranuloma (JXG) is the most common non-Langerhans cell histiocytosis. It is primarily a cutaneous disease that affects infants. To our knowledge only 7 cases of JXG involving the airway have been reported. Our case is the first case of an adult with laryngeal JXG.

Study design: Case report and review of the current literature.

Case report: 22 year old male with large mass involving the left supraglottic larynx. On preliminary biopsy lesion was consistent with high-grade neoplasm likely angiosarcoma. Fully resected with potassium-titanyl-phosphate (KTP) laser without tracheotomy. Final pathology consistent with JXG on histopathology and immunohistochemistry. Three months postoperatively the patient has no recurrence and is speaking, breathing, and eating normally.

Discussion: JXG are rarely seen in extracutaneous sites. They have been reported in the supraglottis, subglottis, and trachea. JXG presents as yellow nodular lesions of the head, neck, and upper trunk.

Conclusions: This is the 1st or 2nd report of these lesions in an adult. These lesions can be removed by KTP laser without the need for a tracheotomy and have normal function postoperatively.

Background

• JXG is a abnormal proliferation of histiocytes that usually presents as a solitary cutaneous lesion within the first year of life
• The natural history of cutaneous JXG is spontaneous resolution with a low chance of relapse of 7%
• Involvement of other organ systems is very rare (4-10%) ; Involvement has been reported in subcutaneous tissue, liver, lung, central nervous system, and spleen
• There are eight reports of airway JGX including this case
• This is the second First report of JGX in an adult

Case Report

• 22 year old male with progressive hoarseness for 1 year
• On exam: large well-defined supraglottic mass involving the right vestibular fold, ventricle, and superior portion of the true vocal fold
• Intraoperative preliminary biopsy: high-grade epithelioid neoplasm
• Laser resection with the KTP laser was preformed
• Overnight hospital stay without need for tracheostomy
• Pathology: 1.4cm yellow with rare spindle shaped cells, multinucleated cells, small histiocyte-like cells, larger lipidized cells, and inflammatory cells
• Immunohistochemistry: Positive for CD68 and negative for S-100 and CD1a.
• Final pathologic diagnosis: Juvenile xanthogranuloma
• Three months postoperatively near normal conversational voice, mobile vocal folds with good pliability; excision site well healed without recurrence of tumor

Table 1: Past report airway xanthogranulomas.

<table>
<thead>
<tr>
<th>Age</th>
<th>Site of lesion</th>
<th>Management</th>
<th>Total length of follow-up</th>
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<tbody>
<tr>
<td>19 weeks</td>
<td>Supraglottis</td>
<td>Tracheal ligation and laser excision</td>
<td>11 months</td>
</tr>
<tr>
<td>24 years</td>
<td>Trachea and bronchus</td>
<td>Tracheotomy</td>
<td>37 months</td>
</tr>
<tr>
<td>3 years</td>
<td>Subglottis</td>
<td>Laser excision</td>
<td>Unreported</td>
</tr>
<tr>
<td>18 months</td>
<td>Tracheal, supraglottis</td>
<td>Laser excision</td>
<td>12 months after tracheotomy resection</td>
</tr>
<tr>
<td>8 years</td>
<td>Trachea</td>
<td>Surgical debulking</td>
<td>Unreported</td>
</tr>
<tr>
<td>5 months</td>
<td>Supraglottis</td>
<td>Laser excision</td>
<td>17 months</td>
</tr>
<tr>
<td>9 months</td>
<td>Supraglottis, glottis, and subglottic</td>
<td>Laser excision</td>
<td>Unreported</td>
</tr>
<tr>
<td>22 years</td>
<td>Supraglottis</td>
<td>Laser excision</td>
<td>3 months</td>
</tr>
</tbody>
</table>

Figure 1: Intraoperative picture of JXG involving supraglottis, glottis and subglottis
Figure 2: Intraoperative photo of KTP laser resection preserving lamina propria; negative margins
Figure 3: Lobular architectural pattern composed of epithelioid, multinucleated cells, plasmacytoid shaped cells with prominent nucleoli
Figure 4: Small histiocytic, larger lipidized cells, and inflammatory cells

References