Juvenile Xanthogranuloma of the Airway

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INTRODUCTION

Juvenile Xanthogranuloma (JXG) is a rare cause of airway obstruction. It is the most common form of non-Langerhans cell histiocytosis, but it characteristically presents as a red-to-yellow, papular, cutaneous lesion (1.2). The usual clinical course of JXG is relatively benign, consisting of lesions that resolve spontaneously within a time frame ranging from one to six years after presentation. We present the Texas Children’s Hospital experience with airway JXG in addition to a literature review for the purpose of providing treatment recommendations.

METHODS AND MATERIALS

The Pubmed database was queried for “Juvenile Xanthogranuloma,” and the words “nose,” “nasal cavity,” “palate,” “tongue,” “pharynx,” “larynx” and “glottis” were used to modify the search. Inclusion criteria were applied to omit cases without documented follow-up, cases without available translation into the English language and cases where disseminated disease was present. One patient with a tracheal lesion was selected from our experience at Texas Children’s Hospital (TCH). Data from all of the cases meeting inclusion criteria was compiled and analyzed in the results section.

RESULTS

Eleven total patients were included in this analysis. The average age at presentation was 75.4 months (median = 72 months). Seven patients were male (63.6%), and four patients were female (36.4%). Lesions were located in the nasal vestibule (9.1%), nasal cavity (9.1%), oral cavity (27.3%), base of tongue (9.1%), supraglottic larynx (9.1%), subglottic larynx (18.2%) and trachea (18.2%) (Figure 1). In patients with laryngeal or tracheal lesions (Figure 2), 100% presented in respiratory distress with an average of 13 weeks of prior noisy breathing noted (median = 6 months). The patient with disease in the base of tongue presented with bleeding from the lesion, and the patients with oral cavity lesions presented with complaints of a painless mass. The patients with nasal lesions presented with complaints of unilateral nasal obstruction (50%) or a persistent, painless mass (50%).

Nine patients (81.8%), including the TCH patient, were initially treated with open or endoscopic surgical excision or debulking. In this group, three patients (27.3%) experienced re-growth of clinically significant disease at an average of 38.7 post-operative weeks (median = 28 weeks). Following re-excision or debulking, no recurrences were reported. In patients who underwent surgical excision or debulking and did not experience recurrence of disease, the average length of follow-up was 23.8 months (median of 22 months). Two patients were initially treated with tracheotomy and observation. In both of these patients, decannulation occurred at an average of 19.5 post-operative months. No deaths or peri-operative complications were reported.

CONCLUSIONS

The spontaneous regression of most types of JXG lesions and low recurrence rates following excision suggest that surgical removal or debulking with close follow-up can be an effective management strategy. Based on our experience and a review of the literature, initial surgical debulking and excision may obviate the need for a tracheostomy. However, tracheostomy should always be considered as a life-saving measure in patients who present in respiratory distress.

REFERENCES