Juvenile Xanthogranuloma of Subglottis: Rare, Recurrent and Refractory

Juvenile xanthogranuloma is a benign, proliferative disorder seen in early childhood. It is the most common form of non-Langerhans cell histiocytosis (LCH) and usually presents with self-resolving cutaneous lesions [1]. Laryngeal xanthogranuloma is extremely rare and only seven cases have been reported in literature. These cases presented with airway obstruction requiring either surgical excision or tracheostomy. Recurrences are not uncommon [2]. A 4-year-old girl presented with acute onset stridor and respiratory distress. She was initially managed as croup with no improvement. On examination, there were no cutaneous lesions, organomegaly or any other abnormality. Contrast-enhanced computed tomography (CECT) neck revealed a well-defined uniformly enhancing tracheal mass at C5-C6 level with marked narrowing of subglottic airway (**Fig. 1)**. Bronchoscopy showed subglottic mass causing significant airway obstruction. Histopathological examination of the mass showed sheets of foamy histiocytic, numerous multinucleated giant cells, plasma cells and areas of hemorrhage. Immunohistochemistry confirmed positivity for CD68 and was negative for CD1a, consistent with the diagnosis of juvenile xanthogranuloma. Following histopathological confirmation, she underwent microlaryngeal surgery. Tracheal mass was excised followed by laser and mitomycin C ablation. She improved and was discharged on short course of steroids.

After five months, she had a recurrence, which was confirmed on bronchoscopy and imaging. She was given steroids and vinblastine as per LCH protocol, but had partial response. On follow-up, child has mild snoring while sleeping; however, there is no respiratory difficulty and she continues to be under close surveillance.

Airway obstruction in young children is usually secondary to foreign bodies or infectious/ inflammatory triggers. Tumorous growths are relatively rare in this age group. Diagnosis in such cases may therefore be delayed. Timely intervention with CT imaging and bronchoscopy is crucial in these scenarios as definitive diagnosis requires histopathological confirmation. Juvenile xanthogranuloma, a benign proliferative disorder of dendritic cell phenotype, is known to have varied biological behaviour from spontaneously regressive cutaneous lesion to rare but fatal central nervous system involvement. The pathogenesis is unknown and the initiating stimuli may be one of many infectious or physical factors [3]. Cutaneous manifestations are most common presenting with solitary yellowish nodules. Extracutaneous manifestations have been reported in 4% of children [4]. Most systemic lesions undergo spontaneous resolution; however, serious complications are known to be associated with few systemic juvenile xanthogranuloma cases [5].

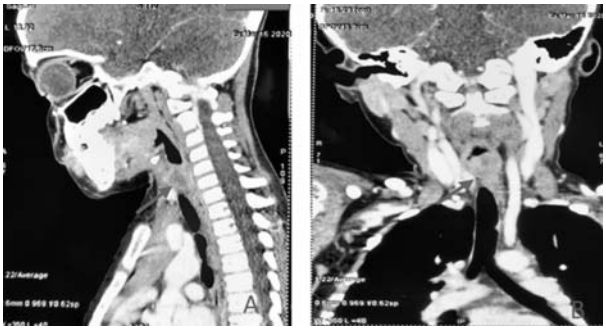

Fig. 1 Contrast-enhanced computed tomography images (sagittal view: A and coronal view: B) showing an elongated mildly enhancing soft tissue mass seen in the glottic and subglottic larynx, extending from the level of posterior commissure at true glottis level and extending inferiorly into the posterior subglottic region, causing marked narrowing of subglottic airway.

Diagnosis of the condition is confirmed by histological features and immunohistochemistry. Treatment of laryngeal juvenile xanthogranuloma includes local excision with or without tracheostomy. In case of recurrence in present case, we achieved partial; though satisfactory, response with steroids and vinblastine as per LCH protocol.

In conclusion, juvenile xanthogranuloma should be considered as a differential diagnosis for subglottic masses causing airway obstruction. Considering its favorable prognosis and tendency of spontaneous regression, decision for reductive surgery with or without tracheostomy needs to be taken judiciously. Close follow-up is advised as recurrence is not uncommon and may require further intervention.

CHANDRIKA VERMA, KUSHAGRA TANEJA,* AMITA MAHAJAN
Department of Pediatric Hematology Oncology, Indraprastha Apollo Hospital, New Delhi.
**kushagrataneja321@gmail.com*

REFERENCES

1. Weitzman S, Jaffe R. Uncommon histiocytic disorders: the non-Langerhans cell histiocytoses. Pediatr Blood Cancer. 2005;45: 256-64.
2. Kawamoto A, Katori Y, Honkura Y, et al. Isolated juvenile xantho-granuloma in the larynx of a three-year-old child. Tohoku J Exp Med. 2013;230:123-7.
3. Chang MW. Update on juvenile xanthogranuloma: Unusual cutaneous and systemic variants. Semin Cutan Med Surg. 1999; 18:195-205.
4. Dehner LP. Juvenile xanthogranulomas in the first two decades of life: A clinicopathologic study of 174 cases with cutaneous and extracutaneous manifestations. Am J Surg Pathol. 2003;27:579-93.
5. Freyer DR, Kennedy R, Bostrom BC, et al. Juvenile xanthogranuloma: Forms of systemic disease and their clinical implications. J Pediatr. 1996;129:227-37.