

Head and Neck Extranodal Rosai-Dorfman Disease- Utility of immunohistochemistry

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Abstract : Background: Rosai-Dorfman disease (RDD), aka sinus histiocytosis with massive lymphadenopathy, is a rare, idiopathic histiocytic proliferative disorder. Although RDD can be seen involving the head and neck lymph nodes, rarely it can affect other extranodal sites. It present 3 unique cases of RDD affecting the nasal cavity, paranasal sinuses, and ear canal. The initial clinical presentation on two cases mimicked a malignant neoplasm. The 3rd case of RDD co-existed with a cholesteatoma of the ear canal. The clinical presentation, histology and immunohistochemical stains, and radiographic findings are discussed. Design: An overview of 3 cases of RDD affected sinonasal cavity and ear canal from UCI Medical Center was conducted. Case 1: A 61 year old male complaining of breathing difficulty presented with bilateral polypoid sinonasal masses and severe nasal obstruction. The masses elevated the nasal floor, and involved the anterior nasal septum to lateral wall. It was endoscopically excised. At intraoperative consultation, frozen section reported a pleomorphic spindle cell neoplasm with scattered large atypical spindle cells, resembling a high grade sarcoma. Case 2: A 46 year old male presented with recurrent bilateral maxillary chronic sinusitis with mass formation, clinically suspicious for malignant lymphoma. Excisional tissue sample showed large irregular spindled histiocytes with abundant granular and vacuolated cytoplasm. Case 3: A 36 year old female with a history of asthma initially presented with left-sided chronic otalgia, occasional nausea, vertigo, and fluctuating pain exacerbated by head movement and temperature changes. CT scan revealed an external auditory canal mass extending to the middle ear, coexisting with a small cholesteatoma. Results: The morphology of all cases revealed large atypical spindled histiocytes resembling fibrohistiocytic or myofibroblastic proliferative neoplasms. Scattered emperipolesis was seen. All 3 cases were confirmed as extranodal sinus RDD, confirmed by immunohistochemistry. The large atypical cells were positive for S100, CD68, and CD163. No evidence for malignancy was identified. Case 3 showed concurrent RDD co-existing with a cholesteatoma. Conclusion: Due to its rarity and variable clinical presentations, the diagnosis of RDD is seldom clinically considered. Extranodal sinus RDD morphologically can be pitfall as mimicker of spindly neoplasm, especially at intraoperative consultation. It can create diagnostic and therapeutic challenges. Correlation of radiological findings with histologic features will help to reach the diagnosis.

Keywords : head and neck, extranodal, ro sai-dorfman disease, mimicker, immunohistochemistry

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