Rosai-Dorfman Disease Presenting As Recurrent Nasal Polyposis

ABSTRACT

Objectives: To present a case of Rosai-Dorfman disease in an individual with a 14-year history of recurrent nasal polyposis and discuss its clinical presentation, physical examination, radiologic findings, histopathologic characteristics and available treatment.

Methods:

Design: Case Report
Setting: Tertiary Government Hospital
Patient: One

Results: A 26-year-old Filipino diagnosed and repeatedly treated medically and surgically for recurrent nasal polyposis underwent repeat endoscopic sinus surgery. A histopathologic impression of Rosai-Dorfman disease was confirmed by positive S-100 and CD1a negative immunohistochemistry in conjunction with the morphologic findings.

Conclusion: Rosai-Dorfman disease is a rare entity which should be considered when dealing with recurrent nasal polyposis that is intractable to initial medical and surgical therapies. Histopathologic findings of emperipolesis and immunohistochemical S-100 stains play a key role in the diagnosis but there is yet no definite treatment for this disease.

Keywords: recurrent nasal polyposis, Rosai-Dorfman disease, emperipolesis, S-100 stain

Nasal polyposis are common pathology of unknown etiology with a high rate of recurrence after surgery. A study of 148 patients to determine the rate and reasons for polyp recurrence after functional endoscopic sinus surgery showed recurrence in 74 patients (49.3%) in the first two years after surgery and 17 patients (12.2%) in the next 3-4 years. There were no associations noted between allergy, infection and recurrence of polyps. Another study that analyzed 10 variables (age, gender, history of purulent nasal discharge, facial pain, anosmia, post nasal drip, headache, nasal allergy, asthma and CT scan staging using the Lund-McKay scoring system) for association with polyp recurrence found a 19% recurrence rate and no association with the variables analyzed except for CT staging which was significantly higher among the group with recurrence compared to the group without recurrence (p<0.00).

Recurrent polyposis which is unresponsive to the standard therapy is associated with hyperplastic chronic sinusitis, allergy, cystic fibrosis and acetylsalicylic (ASA) sensitivity. Patients with ASA sensitivity typically have adult onset asthma with nasal polyps and chronic sinusitis.

Another disease entity associated with recurrent nasal polyposis is inverting papilloma which is a benign but locally aggressive tumor of the nasal cavity. It is similar in appearance but is fleshier than nasal polyp. Histopathology can distinguish the one from the other.

We present a case of recurrent nasal polyposis not responding to repeated medical and surgical treatments diagnosed with Rosai-Dorfman disease.