ABSTRACT

Head and neck lymphomas most commonly occur in the cervical lymph nodes and less frequently in extranodal tonsillar tissue. The paranasal sinuses are an uncommon location for lymphomas and are an exceedingly rare location for Hodgkin lymphoma. Here we present a rare case of a 74 year-old woman with a right maxillary sinus mass with final pathology consistent with Hodgkin's lymphoma and a PET/CT negative for distant disease or nodal involvement. We review the current literature and the proper management and workup of sinonasal masses with clinical concern for a lymphoproliferative process.

CASE PRESENTATION

A 74 year-old nonsmoking Caucasian female with a past medical history including type II diabetes mellitus, hypertension, and congestive heart failure presented with a three week history of right cheek numbness and occipital headaches. Examination revealed hypoesthesia over the right V2 distribution and normal extra-ocular motion. In-office nasal endoscopy revealed purulence draining from the right middle meatus but was otherwise normal. MRI showed a T2-hypointense and T1-isointense enhancing right posterior-lateral maxillary sinus mass involving the infraorbital nerve with questionable involvement of the orbital floor (Figure 1). Subsequent maxillofacial CT scan (Figure 2) confirmed a mass extending into the pterygopalatine fossa with bony involvement including the orbital floor and infraorbital canal.

Patient was taken to the OR and a right maxillary antrostomy and anterior ethmoidectomy was performed to provide access to the large, irregular, and polypoid mass which filled nearly two-thirds of the maxillary sinus. Upon dissection, the mass was adherent to the posterior and lateral walls as well as the roof of the sinus along the infraorbital nerve canal (Fig 3). A small part of the mass located along the infraorbital nerve was left in situ in an effort to avoid paresthesia or neuralgia prior to a known diagnosis of malignancy. Large portions were sent for frozen section, fresh for lymphoma evaluation, and for permanent histopathology. The initial impression at the time of frozen section was a lymphoplasmacytic infiltrate without obvious malignancy, and initial flow cytometric analysis was negative for evidence of a T- or B-cell non-Hodgkin lymphoma. Final H&E sections showed submucosal fibrosis with lymphoplasmacytic inflammation, increased eosinophils, and occasional groups of large, multinucleated, malignant lymphoid cells with prominent nucleoli compatible with Reed-Sternberg cells. Immunohistochemistry confirmed strong, membranous immunoreactivity for CD30 in the Reed-Sternberg cells with immunoreactivity for PAX5; variable immunoreactivity for CD15 and CD20, and minimal immunoreactivity for CD45 (Figure 4). The findings were consistent with mucosal involvement by classical Hodgkin lymphoma, nodular sclerosing subtype.

PET/CT showed no evidence of distant disease or other sites of involvement. She is now undergoing chemotherapy and radiation as per the consensus of the multidisciplinary head and neck tumor board.

DISCUSSION

The majority of head and neck lymphomas occur in the cervical lymph nodes and less frequently in extranodal sites, the most commonly in the tonsillar tissue of Waldeyer's ring. Sinonasal lymphomas represent only 1.5% of all lymphomas, with Hodgkin lymphoma occurring significantly less frequently than non-Hodgkin types, which account for 10 and 90 percent, respectively. Two recent institutional reviews with case series of sinonasal lymphomas reviewed 17 cases over a 22 year period and 23 cases over 38 years, none of which were Hodgkin lymphoma, emphasizing the rare nature of this disease in the sinuses.1,2 Currently, less than five prior case reports of Hodgkin Lymphoma of the paranasal sinuses have been published in the literature.1,4-5 All of these patients underwent similar workups with operative biopsies or resections followed by chemoradiation. A review for the treatment for sinonasal lymphomas showed good initial control with radiation alone, yet a trend for failures with distant recurrence necessitates systemic treatment with chemotherapy.2 Overall survival is strongly correlated with disease stage.

Of paramount importance in this case was the specific testing for lymphoma, which was planned pre-operatively based on the imaging characteristics of the patient’s lesion. Routine frozen and initial flow cytometry were insufficient for accurate diagnosis. Only after combining these studies with final pathology and immunohistochemistry was the diagnosis confirmed.

CONCLUSIONS

Although rare, practitioners must consider the diagnosis of lymphoproliferative diseases for unusual, malignant appearing sinonasal masses. Proper studies, guided by a high index of suspicion, including sending fresh tissue for lymphoma evaluation and flow cytometry in addition to routine histopathology and immunohistochemistry give the best change to obtain a proper diagnosis.

REFERENCES


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