A Unique Presentation of EBV-Associated Castleman’s Disease

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ABSTRACT

Castleman’s Disease, or angiofollicular lymph node hyperplasia, is both a poorly understood benign lymphoproliferative disorder. Castleman’s disease has 2 clinical subtypes:1, unicentric and multicentric and 2 histopathologic types2, hyaline vascular type and plasma cell type. HIV, HHV8, and EBV have been described to be associated with the development of Castleman’s Disease3,4. The role of EBV in the etiology of a Nasopharyngeal Carcinoma (NPC) is well described5. We present a unique case of Castleman’s Disease in a patient, which mimicked the clinical presentation of NPC versus lymphoma, and review the pertinent literature on the topic.

INTRODUCTION

Castleman’s Disease, or angiofollicular lymph node hyperplasia, is both a poorly understood benign lymphoproliferative disorder. Castleman’s disease has 2 clinical subtypes1, unicentric and multicentric and 2 histopathologic types2, hyaline vascular type and plasma cell type. HIV, HHV8, and EBV have been described to be associated with the development of Castleman’s Disease3,4. The role of EBV in the etiology of a Nasopharyngeal Carcinoma (NPC) is well described5. We present a unique case of Castleman’s Disease in a patient, which mimicked the clinical presentation of NPC versus lymphoma, and review the pertinent literature on the topic.

METHODS

A case report and review of the literature.

RESULTS

A 68 year-old male from Southern China presented for evaluation of left level IIa neck mass that was present for 1 year. Biopsy demonstrated reactive changes with follicular hyperplasia. Two years later, the patient returned with multiple enlarged cervical nodes in levels IIa, IIb, and V. Endoscopic and repeat lymph node biopsies revealed only reactive changes with follicular hyperplasia without evidence of malignancy or lymphoma. Three years later, the patient presented with enlarging diffuse left neck and parotid lymphadenopathy with multiple inclusive repeat FNAs. Hematology-Oncology and Infectious Disease consultation were unrevealing and considered more aggressive surgical management necessary to assist in diagnosis. Radiographically, CT scan showed interval increase in the size and number of cervical adenopathy suggestive of metastatic disease (Tables 1 and 2). The patient consequently underwent a left superficial parotidectomy and neck dissection (Figure 3).

Surgical histopathology displayed reactive lymphoid tissue with a morphologic pattern consistent with hyaline-vascular type of Castleman’s Disease with extensive fibrosis (Figure 4). Immunohistochemistry staining was positive for EBV (EBER) with a marked expansion of IgG4 plasma cells suggesting potential association with an IgG4-related sclerosing disease. However, serum IgG4 levels were normal which ruled out this disorder Postoperatively, the patient did well and required no further intervention.

CONCLUSIONS

Castleman’s Disease is a lymphoproliferative disorder that can be associated with EBV, EBV infection may play a role in the progression of Castleman’s Disease in individuals from Southern China, as similarly seen with NPC, and be difficult to diagnose on initial biopsy. Multidisciplinary assessment and re-evaluation is necessary for patients with recurrent reactive lymphadenopathy of unknown etiology.

REFERENCES

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