INTRODUCTION

FDCS is a neoplasm of follicular dendritic cells (FDC), which are non-lymphoid non-phagocytic antigen presenting cells of the reticuloendothelial system. FDCS occurs intra- or extranodally. Intranodal disease most commonly presents in cervical, mediastinal, axillary, and abdominal nodes. Extranodal disease most commonly presents in abdominal organs, head and neck, and mediastinum (1).

In the English language literature on Medline, there are less than 150 cases reported of FDCS in the head and neck, 12 of which are reported in the parapharyngeal space (2).

CASE PRESENTATION

42-year-old male with incidentally noted Poland Syndrome (congenitally absent unilateral pectoralis major with ipsilateral digit deformities), presented with a rapidly enlarging left parapharyngeal tumor who underwent three biopsies at three different institutions with inconclusive diagnoses over 4.5 months. Imaging revealed a 52mm parapharyngeal mass invading into nasopharynx and carotid space, suggesting neurogenic tumor. Fine needle aspiration was obtained, but again non-diagnostic. Further confounding the clinical picture, a pacemaker was required for multiple episodes of sinus pause due to mass effect on the carotid body. Ultimately, patient underwent transpalatal and transcervical approach to the parapharyngeal space and transoral partial glossectomy, palatectomy, and pharyngectomy with reconstruction for diagnostic and definitive resection.

Final pathology resulted as FDCS with immunophenotype of non-neoplastic FDCs, with CD21, CD35 and CD23, and vimentin with a low level of B and T staining and a level of A. To present a case of FDCS, to review the differential diagnosis of FDCS, and to discuss the current management approaches to FDCS.

CONCLUSION

Primary tumors of FDCS were postulated by Lennert in 1978 (3), but was not reported until 1986 by Monda et al. (4). Chan et al. described the first extranodal cases in 1994 (5). Although there are case series and review articles on this neoplasm, it remains under-recognized, with up to 58% of extranodal cases being initially misdiagnosed (6).

FDCS diagnosis is frequently delayed due to misdiagnosis of biopsies containing reactive necrotic and reactive lymphoid. Differential includes thymoma, melanoma, meningioma, undifferentiated carcinoma, lymphoepithelial carcinoma, spindle cell carcinoma, peripheral nerve sheath tumor, sarcomas, and GIST (7).

Mean age is 44 years, with no female or male predilection. The only association is that 10–20% of cases have preceding or concurrent Castleman disease (1,6,7).

Histologically, FDCS appears spindle-like with prominent whorled, storiform pattern, and interdigitating processes. Nuclei are oval or rounded, may be irregular in contour with pleomorphism (1,7). Poor prognosis is based on size >6cm, presence of coagulative necrosis, high mitotic count (>5/10 hpf), nuclear pleomorphism, intra-abdominal location, and lack of adjuvant therapy(7).

FDCS cells share the immunophenotype of non-neoplastic FDCs, with CD21, CD35 and CD23 being the most specific markers. Other markers include vimentin and desmoplakin. Some are variably positive for EMA, S100, and MSA, presenting diagnostic pitfalls (1).

Due to the variable clinical course of FDCS, there is no standardized treatment regimen. FDCS is often treated with wide local resection with or without radiotherapy and/or chemotherapy (CHOP or CHOP-like regimens). One review showed overall recurrence rate of 43%, metastatic rate 24%, and mortality rates 17%. For extranodal FDCS, 2- and 5-year recurrence-free survival rates were 62.3% and 27.4% respectively (7).

REFERENCES