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Case Report

Recurrence of follicular dendritic cell sarcoma in tonsils after 7 years: a rare case report

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ABSTRACT

Follicular dendritic cell sarcoma (FDCS) is a rare mesenchymal neoplasm. It arises from not only from lymph nodes but also from extra nodal tissues, either as acquired lymphoid tissue or as part of the organized constitutive lymphoid tissue. We here report this rare entity which developed again after 7 years post tonsillectomy in a 5-year old male patient. Patient underwent radical tonsillectomy and adjuvant treatment is awaited. Differential diagnosis includes large cell lymphoma, peripheral nerve sheath tumor, extracranial meningioma, malignant melanoma, metastatic carcinoma, ectopic thymoma, malignant fibrous histiocytoma, and interstitial reticulum cell sarcoma. Currently, the management of FDCS includes the therapeutic guidelines similar to that of high-grade soft tissue sarcomas that is complete surgical resection of the lesion with possibility of adjuvant radiotherapy and/or chemotherapy. The ideal combination of management of FDCS has yet to be defined.

Keywords: Follicular dendritic cell sarcoma, FDCS, Oropharyngeal cancer, Tonsillar mass, Radical tonsillectomy

INTRODUCTION

Follicular dendritic cell sarcoma (FDCS) was first described by Monda et al in 1986 in a report of four case of non- lymphomatous primary lymph node malignancy.[1] It arises from not only from lymph nodes but also from extra nodal tissues, either as acquired lymphoid tissue or as part of the organized constitutive lymphoid tissue.[2] World Health Organization (WHO) classified FDCS into histocytic and dendritic cell neoplasm which includes histocytic sarcoma, Langerhan cell tumors and interdigitating dendritic cell tumors/ sarcomas.[1,3]

Uncontrolled proliferation of follicular dendritic cells which are accessory immune cells affects lymph nodes, mediastinum and cervical and axillary region. When it involves extra-nodal region, pharyngeal region is one of the preferred sites.[4] FDCS has clinical and histologic similarity with p16-positive human papillomavirus (HPV)-related squamous cell carcinoma of the oropharynx which characteristically has non-keratinizing morphology and frequently presents as an isolated neck mass.[5] We, here report this rare entity which developed again after 7 years post tonsillectomy in a 54-year-old male patient. Patient underwent radical tonsillectomy and adjuvant treatment is awaited.

CASE REPORT

A 54-year-old male presented with chief complaint of difficulty in swallowing since 1 month. It was not associated with hoarseness and otalgia.

He had history of left tonsillectomy in 2015 and on histopathology was highly suspicious of sarcomatoid carcinoma and lympho-epithelioma. Further IHC marker studies were performed and the tumor cells expressed suggested CD 23, CD 21 (focal), CD 35 and fascin. The tumor cells were immune-negative for cytokeratin, CD 20

and CD 3 and concluded follicular dendritic cell sarcoma as the final diagnosis.

On Hopkins examination - a nodular swelling was seen in left lateral wall of region pharynx and tonsillar region. On computed tomography (CT) neck with contrast (June 22), an ill-defined soft tissue density with heterogeneous enhancement measuring 25×14 mm noted arising from postero-lateral wall of oropharynx. Few sub-centimetric lymph nodes seen with largest measuring 12×7 mm in level II on left side.

On magnetic resonance imaging (MRI) neck with contrast (July 22) the left palatine tonsil appeared diffusely bulky with heterogeneous morphology with avid enhancement on post contrast acquisition likely to be recurrence and lymph node showed heterogenous enhancement suggested early necrosis along level II, measures 6-8 mm in short axis diameter.

Radical tonsillectomy with SOHND was performed and specimen sent for histopathology and IHC (Figures 1a and b). Sections showed tonsillar tissue with tumor cells where most of the tumor had spindloid cells arranged in storiform pattern and some had histiocytic appearance. Lymphocytic cell collection and few mitotic cells were seen highly suggestive of sarcomatoid carcinoma. On immunohistochemistry examination tumor cells were positive for CD 23, CD 21 (focal), CD 35 and fascin. Thus, final diagnosis came out to be FDSC.

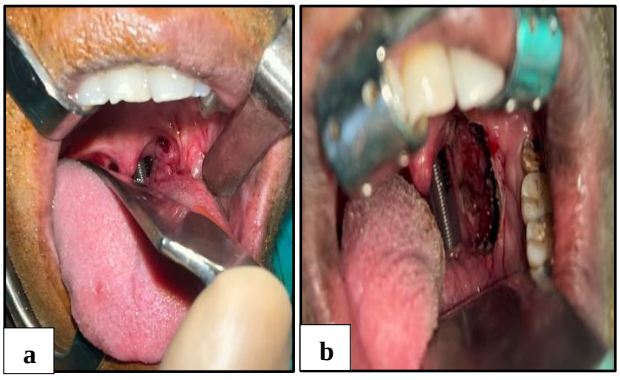

Figure 1: (a) Recurrent lesion present over left tonsillar region before surgical excision, and (b) surgical defect following radical tonsillectomy.

DISCUSSION

FDCS is extremely rare which was first described by Monda et al in 1986 and very few such cases involving the extra-nodal sites have been reported worldwide.[1,2] Duan et al described FDCSs involving soft palate, retromolar trigone, nasopharynx and parapharyngeal space.[6] Zhou et al reported a single case arising from the parotid.[7] Chan et al described the first case of FDCS of palate which was earlier misdiagnosed as an acinic cell carcinoma.[8]

Etiology and pathogenesis for FDCS is still not clear but certain cytogenetic abnormalities in the spleen with multiple unbalanced clonal chromosomal translocations including loss of Xp were described in one of the case.[9] This can be early evidence that Xp might play an important role in development of tumour. The transformation may be due to FDC dysplasia in CD follicles following the sequence of hyperplasia then dysplasia and neoplasia like development of few epithelial neoplasms and this forms basis of FDCS formation.[10,16]

The transformation of p53 tumour suppressor with overexpression of p53 protein and increased p53 positive spindle cells in hyaline vascular CD specimen were also noted in FDCS.[10] In some cases Ebstein Barr virus was seen positive and its association with it as FDCS expresses EBV receptor (CD21) and so infected by it. Pathogenesis of FDCS due to EBV is unclear but LMP-1 (latent membrane protein-1) is encoded by LMP-1 gene and is an integral membrane protein of EBV and considered as viral oncogene that transforms rodent fibroblasts and render them tumorogenic in mice in vitro.[11]

FDCS appears as a well-circumscribed, solitary, solid mass with white, pink or grayish tan color. Usually tumor cells are ovoid, spindled or polygonal in shape with an eosinophilic cytoplasm, and indistinct borders, ensuing a syncytial appearance. The nuclei appear to be oval to round in shape with smooth borders and mild atypia. Presence of small lymphocytes intermixed with tumor cells around the blood vessels (perivascular cuffing) is a characteristic histological feature seen. Often the microscopic architecture resembles that of sarcomas as fascicular, storiform, whorled, diffused, follicle like or trabecular. There is variation in the mitotic rate, but it is usually low to intermediate.[12,13] Also, the tumor necrosis is uncommon.

Lan et al. proposed a prognostic assessment system for FDCS. Tumor size ≥5 cm, mitotic count ≥5/10 HPF and high-grade histology were related to tumor recurrence. Depending upon five histological parameters (architecture, cellular features, mitotic count, necrosis and lymphocytic infiltration) and one immuno-histochemical parameter (Ki-67 proliferative index), they divided FDCS into low-grade and high-grade varieties.[9]

The neoplastic cells in FDCS generally shows the immune-phenotype of non-neoplastic follicular dendritic cells and are usually positive for CD21, CD23 or CD35. They are also generally positive for desmoplakin, fascin, vimentin, human leukocyte antigen-DR and EMA, and are variably positive for CD68 and S-100.[10,16] The expression of CD1a, desmin and CD45 were not seen in FDCS and hence can be differentiated from Langerhans cell tumors, interdigiting dendritic cell tumors, and lymphoid and histiocytic neoplasias. Hence, expression of these non-typical markers should be considered for differential diagnosis with other neoplasms.[15]

Li et al in a review of 137 cases of FDCS in head and neck region found, 72 cases of extranodal FDCS and 65 cases of nodal FDCS where for extranodal cases, 33 (45.8%) were in palatine tonsil, 11 (15.3%) in nasopharynx, 8 (11.1%) in parapharyngeal space, 6 (8.3%) in soft and hard palate, 3 (4.2%) in thyroids, 3 (4.2%) in parotid gland and 8 (11.1%) at other sites. For nodal FDCS cases, 20 (30.8%) were in right cervical lymph node, 21 (32.3%) in left cervical lymph node, and 24 in undetermined cases.[15] Differential diagnosis includes large cell lymphoma, peripheral nerve sheath tumor, extracranial meningioma, malignant melanoma, metastatic carcinoma, ectopic thymoma, malignant fibrous histiocytoma, and interstitial reticulum cell sarcoma.[16-18] Due to its rare occurrence, FDCS is hardly in mind as a differential diagnosis primarily, and its clinical pattern and most advantageous management has not been consolidated.

Currently, the management of FDCS includes the therapeutic guidelines similar to that of high-grade soft tissue sarcomas that is complete surgical resection of the lesion with possibility of adjuvant radiotherapy and/or chemotherapy. The ideal combination of management of FDCS has yet to be defined.[15]

CONCLUSION

Extra-nodal FDCS of the pharyngeal region is rare entity in head and neck region and can be misdiagnosed at higher rates. So, in order avoid this FDCS should always be considered as differential diagnosis for any tonsillar mass. Histo-pathological examination with IHC is most important to conclude the diagnosis as CD21 and CD35 antibodies remain positive in FDCS. Due to its rarity in occurrence further research is required to implement standard guidelines for its management and prevent recurrences.

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REFERENCES

1. Monda L, Warnke R, Rosai J. A primary lymph node malignancy with features suggestive of dendritic reticulum cell differentiation. A report of 4 cases. Am J Pathol. 1986;122:562-72.
2. Eun YG, Kim SW, Kwon KH. Follicular dendritic cell sarcoma of the tonsil. Yonsei Med J. 2010;51(4):602-4.
3. Jaffe ES, Harris NL, Stein H, Vardiman JW. Tumours of Haematopoietic and Lymphoid Tissues. Lyon: IARC. 2001.
4. Duan G, Wu F, Zhu J, Guo D, Zhang R. Extranodal follicular dendritic cell sarcoma of the pharyngeal region. Am J Clin Pathol. 2010;133(1):49-58.
5. Zhang L, Yang C, Lewis JS Jr, El-Mofty SK, Chernock RD. p16 expression in follicular dendritic cell sarcoma: a potential mimicker of human papillomavirus-related oropharyngeal squamous cell carcinoma. Hum Pathol. 2017;66:40-7.
6. Duan GJ, Wu F, Zhu J, Guo D, Zhang R, Shen L, et al. Extranodal follicular dendritic cell sarcoma of the pharyngeal region. A potential diagnostic pitfall with literature review. Am J Clin Pathol. 2010;133:49-58.
7. Zhou Z, Zheng LY, Wang JD, Liu Q. Follicular dendritic cell sarcoma of parotid gland: 1 case report. Chin Arch Otolaryngol Head Neck Surg. 2005;12:138.
8. Chan JK, Tsang WY, Ng CS, Tang SK, Yu HC, Lee AW. Follicular dendritic cell tumors of the oral cavity. Am J Surg Pathol. 1994;18:148-57.
9. Sander B, Middel P, Gunawan B, Schulten HJ, Baum F, Golas MM, et al. Follicular dendritic cell sarcoma of the spleen. Hum Pathol. 2007;38(4):668-72.
10. Chan AC, Chan KW, Chan JK, Au WY, Ho WK, Ng WM. Development of follicular dendritic cell sarcoma in hyalinevascular Castleman's disease of the nasopharynx: tracing its evolution by sequential biopsies. Histopathology. 2001;38(6):510-8.
11. Wang D, Liebowitz D, Kieff E. An EBV membrane protein expressed in immortalized lymphocytes transforms established rodent cells. Cell. 1985;43:831-40.
12. Perez-Ordonez B, Erlandson RA, Rosai J. Follicular dendritic cell tumor: report of 13 additional cases of a distinctive entity. Am J Surg Pathol. 1996;20(8):944-55.
13. Duan GJ, Wu F, Zhu J, Guo DY, Zhang R, Shen LL, et al. Extranodal follicular dendritic cell sarcoma of the pharyngeal region: a potential diagnostic pitfall, with literature review. Am J Clin Pathol. 2010;133(1):49-58.
14. Li L, Shi YH, Guo ZJ, Qiu T, Guo L, Yang HY, et al. Clinicopathological features and prognosis assessment of extranodal follicular dendritic cell sarcoma. World J Gastroenterol. 2010;16:2504-19.
15. Li J, Zhou ML, Zhou SH. Clinical and pathological features of head and neck follicular dendritic cell sarcoma. Hematology. 2015;20(10):571-83.
16. Soriano AO, Thompson MA, Admirand JH, Fayad LE, Rodriguez AM, Romaguera JE, et al. Follicular dendritic cell sarcoma: a report of 14 cases and a review of the literature. Am J Hematol. 2007;82(8):725-8.
17. Domínguez-Malagón H, Cano-Valdez AM, Mosqueda-Taylor A, Hes O. Follicular dendritic cell sarcoma of the pharyngeal region:histologic, cytologic, immunohistochemical, and ultrastructural study of three cases. Ann Diagn Pathol. 2004;8:325-32.
18. Aydin E, Ozluoglu LN, Demirhan B, Arikan U. Follicular dendritic cell sarcoma of the tonsil: case report. Eur Arch Otorhinolaryngol. 2006;263:1155-7.

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