**INTRODUCTION**

Follicular dendritic cells like T-lymphocytes, B-lymphocytes, macrophages and N.K cells are among the accessory cells in the immune system. They are present in the germinal centres of lymphoid follicles in the spleen and lymph nodes. They have numerous dendritic cytoplasmic process. They bear Fc receptors for Ig G so they can trap antigen bound to antibodies and present them to B cells. The cells express positivity towards CD21, CD 35 and S-100 monoclonal antibodies. Follicular dendritic cell sarcoma (FDCS) is unusual and its extranodal origin is extremely rare. Seventy one cases have been reported in the English language medical literature, of which 25 cases were extranodal, occurring in the head and neck region. Twelve cases of tonsillar FDCS have been reported.

We report a new case of FDCS of tonsil in a 52 years old woman.

**CASE REPORT**

The patient was a 52-year-old woman with no comorbidity. She presented to the out patient department with complaints of a swelling in the throat and dysphagia for the last few weeks. There was no history of fever, otalgia or trismus. On clinical examination, the right tonsil was found to be enlarged and indurated with a normal mucus membrane covering. The base line investigations were normal. CT scan of the head and neck area, ultrasound of abdomen and Mountoux test were all unremarkable. Diagnostic tonsillectomy was performed and a specimen of size 2.5x2 cm was sent for histopathology. Preliminary diagnosis was suggestive of “non-epithelial malignant neoplasm”. Histological sections revealed proliferation of spindled to ovoid cells. The tumour cells had eosinophilic cytoplasm with indistinct cell borders. The nuclei were elongated with vesicular to finely dispersed chromatin (Figure 1). There was mild to moderate nuclear atypia and pleomorphism. Mitotic activity was enhanced. In some areas lymphocytes were intermixed with neoplastic cells. Overlying stratified squamous epithelium was unremarkable. The sections were stained with a pannel of monoclonal antibodies using "Envision system”. The tumor cells expressed CD21 and S-100 protein. CD6 B was equivocal on additional immunohistochemical stain.

The final diagnosis was FDCS with negative margins of resection. Skeletal scintography was found negative for metastatic involvement of the skeleton. Chemotherapy
(Doxorubicin and Ifosfamide) was given with consultation of the oncologist. The patient was free of disease one year after the end of the treatment.

**DISCUSSION**

FDC sarcoma is a rare tumour that derives from the dendritic cells of lymphoid follicles. Follicular dendritic cells are non-lymphoid, non-phagocytic, accessory cells in the immune system that are essential for antigen presentation and germinal centre regulation. Lymph nodes are the sites most commonly involved by FDC sarcoma and its extranodal origin is extremely rare. However, it may arise at a variety of extranodal sites including the oral cavity, tonsils, gastroi and liver, because of the presence of dendritic cells there.

Only a few cases of FDCS of the head and neck region have been reported. It occurs most frequently in adults aged between 30 and 50 years, with no gender predilection. The maximum age was noted to be 77 years in a woman, who was previously misdiagnosed as squamous cell carcinoma of the tonsil.

Histologically, tumour cells present with abundant eosinophilic cytoplasm, hyperchromic and pleomorphic nuclei and prominent nucleoli. Immunohistochemically, tumour cells were strongly and diffusely positive for follicular dendritic cells markers CD21, CD35, CD1a, S100 protein. These markers are, therefore, essential for correct diagnosis.

The extranodal tumours, therefore, can easily be misdiagnosed as FDC markers and are not routinely used in the immunohistochemical study of poorly differentiated tumours. Differential diagnosis is to be made from several tumours, including undifferentiated carcinoma, squamous cells carcinoma, malignant melanoma, large cell lymphoma, menengioma, thymoma, malignant fibrous histiocytoma, peripheral never sheath tumour, angiofibrosarcoma and inflammatory pseudotumours.

Treatment modalites include surgery followed by radiotherapy or chemotherapy or an adjuvant chemoradiotherapy. However, surgery is the primary treatment and a wide resection is recommended when the diagnosis has been established pre-operatively in cases with postoperative positive margins and in recurrent cases. Neck dissection in a clinically negative neck examination is controversial and there is no consensus opinion described in the previously reported cases.

However, neck dissection is recommended in cases with evidence of cervical node metastasis. In this case, neck dissection was not done as CT showed no evidence of cervical node metastasis. In the 12 reported cases of tonsilar FDCS, only 3 patients had lymph node metastasis but 5 patients underwent neck dissection.

The role of postoperative adjuvant treatment is also debatable. Some authors suggest systemic radiotherapy in all the resected cases, while others reserve this option in neck node metastasis, cases with positive resected margins, tumours with adverse pathological features and in recurrent cases. A one year follow-up in this case showed no evidence of regional recurrence. Aydin et al. reported the case of a 76 years old female of FDCS of tonsil who was disease free at 4 years follow up. Ludwig reported the case of a 47 years old female who had 3 recurrences within a period of 11 years and the recurrences showed a more aggressive FDCS behaviour than was initially assessed. Due to the limited number of cases reported with short follow-up, a general consensus about the significance of neck dissection, prognostic role of adjuvant therapy and frequency of recurrence has not been established.

**REFERENCES**