Follicular dendritic cells are the stromal cells that are present in the germinal centers of the lymphoid tissue. They trap antigens on their surface and then present them to the B cells. Follicular dendritic cell sarcoma (FDCS) arising in the lymph nodes was first reported by Monda et al. in 1986. FDCSs often arise in lymph nodes, although the extranodal presentations have also been described.

It is difficult to make a diagnosis of FDCS with a fine-needle aspiration cytology. In addition, it is not generally included in the differential diagnosis of tumors that are composed of spindle and ovoid cells with scattered mature lymphocytes in the head and neck region. Initially, these features suggest a wide variety of other diagnoses such as carcinoma, malignant fibrous histiocytoma, schwannoma and ectopic meningioma. In cases of FDCS, the tumor cells show immunoreactivity for the markers specific for follicular dendritic cell differentiation and these include CD21, CD23, and CD35. Combined with characteristic histological features, these immunohistochemical findings allow diagnosis of FDCS. 

Primary treatment of FDCS is surgical excision with or without adjuvant therapies, such as radiotherapy or chemotherapy. FDCS usually shows an indolent clinical course like other low-grade soft tissue sarcomas. It has recently been reported that the recurrence rate of FDCS can be as high as 50%.

Follicular dendritic cell sarcoma (FDCS) is a rare malignancy arising from the antigen-presenting cells in the lymph node and extranodal tissue. We describe a 31-year-old male patient who presented with a swelling of the left parapharynx. The radiologic findings showed a 4.7×4.5×1.9 cm-sized, ill-defined mass in the left parapharyngeal space. A fine-needle aspiration cytology was performed and it showed scattered, irregular, cohesive clusters of tumor cells with a spindle-to-ovoid shape with irregular contours in a background of lymphocytes. Based on these findings, a diagnosis of spindle cell neoplasm was made. The surgically resected tumor was composed of elongated, ovoid or polygonal cells showing positive immunohistochemistry for CD21, CD23, and CD35. Postoperatively, the residual tumor was observed to undergo a rapidly growth. There is an overlap in the cytologic and histologic findings between FDCS of the parapharynx and other tumors. Pathologists should therefore be aware of its characteristics not only to provide an accurate diagnosis but also to recommend the appropriate clinical management.

Key Words: Dendritic cell sarcoma, follicular; Extranodal; Parapharynx

Received: January 28, 2012
Revised: March 28, 2012
Accepted: March 29, 2012

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Extranodal Follicular Dendritic Cell Sarcoma

The occurrence of extranodal FDCS is associated with a larger size (≥ 5 cm), a high-grade histology and a high mitotic count (≥ 5/10 high power fields [HPFs]). However, the precise clinical behaviors and biologic features of this malignancy in the head and neck region remain unknown. We herein report a case of FDCS in the parapharyngeal space which was preoperatively diagnosed using a fine-needle.

CASE REPORT

A 31-year-old man presented with a swelling of the left parapharynx. Physical examination revealed a hard, non-tender mass with round shape in left posterior pharyngeal area with no enlarged lymph nodes in the neck.

On initial computed tomography (CT) scans, there were an ill-defined, heterogeneous mass in the left parapharyngeal space but no remarkable enlarged lymph nodes in the neck (Fig. 1A). On magnetic resonance image scans, there were a 4.7 × 4.5 × 1.9 cm-sized lobulated enhancing mass with a hyperintensity on T2WI (Fig. 1B) and an isointensity on T1WI. The impression of radiologists was neurogenic tumor or pleomorphic adenoma. On positron emission tomography-CT scans, the mass of left parapharyngeal area showed a focal hypermetabolic lesion (SUV = 6.1) (Fig. 1C).

On the fine-needle aspiration cytology, there was a high cellular smear. The scattered, irregular, cohesive clusters of tumor cells showed irregular contours with mature lymphocytes scattered in the background (Fig. 2A). The spindle-to-ovoid tumor cells had an indistinctive cell border and a moderate amount of pale eosinophilic cytoplasm. In tumor cells, oval-shaped nuclei showed mild atypia with a fine chromatin pattern and they occasionally had small distinctive nucleoli (Fig. 2B). Nuclear pseudoinclusions were seen in some tumor cells (Fig. 2B inset). We made a diagnosis of a spindle cell neoplasm including paraganglioma.

Grossly, the surgically resected specimen showed a fibrotic cut surface with a grayish white color (Fig. 3A). Histopathologically, there was a complete replacement of the normal tissue by...
the tumor. The tumor was composed of the elongated, ovoid or polygonal cells and it exhibited a fascicular growth pattern in certain areas. The tumor cells had more or less abundant eosinophilic cytoplasm with ill-defined cell borders. They had not only round-to-oval nuclei with a smooth nuclear membrane but also vesicular or granular chromatin and distinct nucleoli. In addition, they also had a nuclear pleomorphism with a sparse mitotic count (0-1/10 HPFs). Small lymphocytes were scattered throughout the tumor (Fig. 3B). Hemorrhages, but not necrosis, were detected in the peripheral area.

On immunohistochemistry, the tumor cells were positive for CD21, CD23, and CD35, all of which are markers specific for follicular dendritic cell differentiation (Fig. 3C-E). However, the tumor cells were negative for pancytokeratin (AE1/3), cytokeratin 19, human melanoma black-45, chromogranin, synaptophysin, CD56, leukocyte common antigen, smooth muscle...
actin, desmin, and S-100 protein. The ki-67 labeling index was about 10%.

In the current case, a diagnosis of FDCS of the parapharynx was therefore confirmed, based on the histologic and immunohistochemical findings.

A follow-up CT scan was performed on postoperative day 19, and it revealed a peripheral enhancing lesion with an ill-defined margin on the inferior side of the primary lesion (Fig. 1D). These findings are suggestive of the residual tumor, that was rapidly grew. The tumor extend that its greatest dimension reached 3 cm in the short-term postoperative period. The patient underwent the postoperative adjuvant radiotherapy.

**DISCUSSION**

FDCS typically arises in lymph nodes of the neck, axilla and mediastinum, and it also occurs less commonly in extranodal tissues from variable sites, including head and neck, liver, spleen, gastrointestinal tract, soft tissue, skin, lung, and breast.

According to a review of English literatures, six cases of FDCS arising in the parapharyngeal space have recently been described. But there is no case reported in Korea. The clinical data of previously reported six cases of FDCS of the parapharynx are summarized in Table 1. Most cases of FDCS of the parapharynx showed a more aggressive behavior with a local recurrence or a distant metastasis. There were two cases of FDCS of the parapharynx with the greatest diameter of <5 cm, one of which underwent no adjuvant therapy and had a local recurrence. Most cases of FDCS of the parapharynx showed a more aggressive behavior with a local recurrence or a distant metastasis as compared with that of other regions. The current case showed a low mitotic count and a small-sized mass, indicating a better prognosis, but it did an aggressive behavior with a local recurrence and a rapid growth.

It is therefore necessary to examine the pathogenesis of FDCS, which would be essential for predicting a prognosis and selecting the optimal treatment modalities. There is an overlap in the cytological and histologic findings between FDCS and other tumors. Misdiagnosis can be avoided, however, if pathologists are more aware of its characteristic features.

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**Table 1. Summary of six cases of extranodal FDCS of the parapharyngeal region that have previously been reported in the literature**

| Reference            | Sex/ Age (yr) | Size (cm) | Presentation of cytologic features | Initial diagnosis | Applied treatments | Recurrence                  |
|----------------------|---------------|-----------|-----------------------------------|------------------|--------------------|-----------------------------|
| Chan et al. (1997)   | F/40          | 7 x 3 x 2 | No                                | NA               | Excision           | Local recur at 1 yr         |
| Desai et al. (1999)  | F/45          | 6 x 3 x 3 | No                                | Ectopic meningioma | Excision           | Local recur at 1 yr with lymphatic embolization and angionvasion |
| Vargas et al. (2002) | F/54          | 6         | No                                | FDC tumor        | Excision and radiotherapy | Recur at 6 mo in skull base |
| Satoh et al. (2003)  | M/16          | 3 x 2.5   | No                                | Low-grade maclignant tumor | Excision and chemo- and radiotherapy | No recur |
| Dominguez-Malagón et al. (2004) | M/26 (large mass) | NA | No | FDCS | Subtotal resection | Combination of radio- and chemotherapy after distant metastasis | Multiple metastasis in lungs |
| Alexander et al. (2007) | M/69 | 3 | No | Paraganglioma | Excision | Radiotherapy after recurrence | Recur at 1 yr |

FDCS, follicular dendritic cell sarcoma; F, female; NA, not available; FDC, follicular dendritic cell.
Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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