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Case Report

A subcutaneous follicular dendritic cell sarcoma occurring in the cheek: case report

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Abstract: We report a subcutaneous follicular dendritic cell sarcoma (FDCS) in the cheek. Follicular dendritic cell sarcoma is a rare neoplasm. Here we report the case of an 81-year-old woman presenting with the chief complaint of swelling of the left cheek. We could only diagnose granulation tissue based on a core-needle biopsy; thus, we decided to excise the tissue for definitive diagnosis. At first, we considered a B-cell lymphoma, but the diagnosis of FDCS was made following immunostaining of FDC markers. Tumor resection was performed without any other treatments such as chemoradiotherapy. No recurrence has so far been detected for three years. In the head and neck, FDCS occurs with a predilection for the tonsils. This appears to be the first report of a case of FDCS occurring subcutaneously in the cheek. Currently, there are several options for treatment and can only be standardized by accumulating experience with future cases.

Keywords: Follicular dendritic cell sarcoma, head and neck, subcutaneous, immunohistochemical staining

Introduction

Follicular dendritic cells (FDC) are antigen-presenting cells found in lymphoid follicles that help maintain the lymph-node structure. FDC sarcomas (FDCS) are classified as histiocytic and dendritic cell neoplasms by the World Health Organization and are believed to originate from FDC. Although they mainly occur in lymph nodes of the neck and the abdominal cavity, majority of the cases of the head and neck FDCS arise in the tonsils (**Table 1**). We report a case in which a subcutaneous FDCS developed in the cheek.

Case report

An 81-year-old woman presented with swelling of the left cheek. Blood tests revealed no abnormalities; T1-weighted magnetic resonance imaging (MRI) revealed low signals and T2-weighted MRI revealed a mixture of low and high signals (**Figure 1**). A firm and partially mobile tumor was palpable. The tumor was diagnosed following a core-needle biopsy, but because a definitive diagnosis could not be made, tumor excision was planned.

During the surgery, an incision under the canine confirmed the presence of the tumor, and because it surrounded the infraorbital nerve, it appeared to be an infraorbital nerve schwannoma. We removed the entire infraorbital nerve, including the surrounding tissues. There was no tumor infiltration in the surrounding tissues, and the tumor was dissectible from the surrounding tissues (**Figure 2**).

Histopathology

Immunohistochemical staining revealed numerous B lymphocytes expressing CD20 and CD10 with few atypical cells in addition to $CD3^+$ and $CD4^+$ T lymphocytes. We initially considered low-grade B-cell lymphoma [1]. However, HE staining revealed a solid tumor with some whorl formations comprising spindle-shaped and ovoid cells. Immunohistochemical staining was positive for 4 FDC markers, including CD21, CD23, CD35, and CAN.42, and CD79a, and, on the other hand, negative for S-100, α-SMA,

Table 1. Head and neck FDCS reported in literatures [3-5]

Site	Number of cases
Tonsil	62
Parapharyngeal space	4
Soft palate	2
Nasopharynx	6
Hypopharynx	1
Cheek (present case)	1

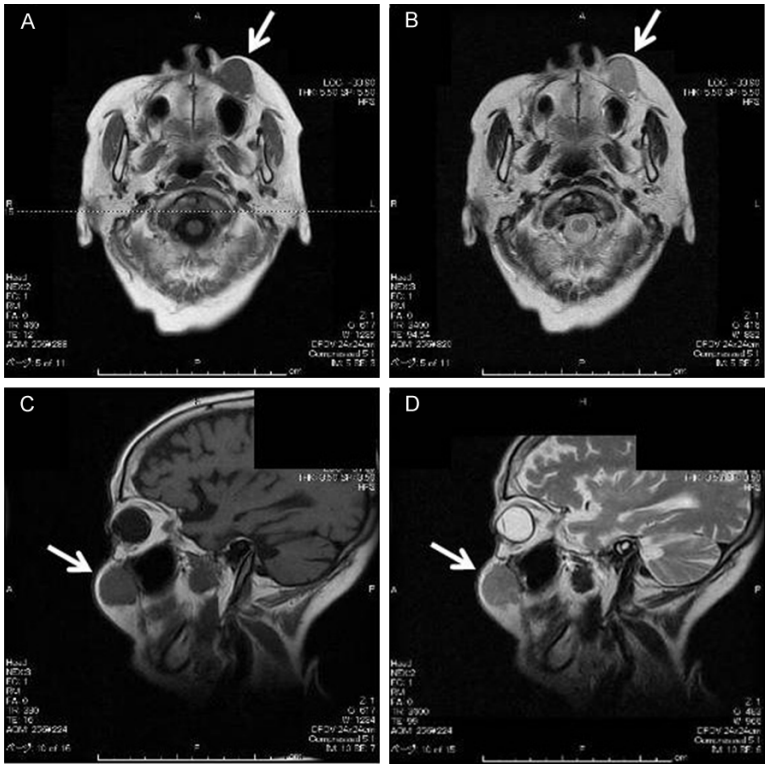

Figure 1. MRI imaging of the tumor (arrow): MRI showing low-intensity signals in T1-weighted (A and C) and mixture of low-intensity and high-intensity signals in T2-weighted images (B and D).

CD3, EGFR, and ALK-1 (**Figure 3**). In addition, EBER-1 RNA was not expressed as revealed by *in situ* hybridization. The diagnosis was subcutaneous FDCS in the cheek. The MIB-1 index was 70%, exclusive of non-tumor cells, in hot spots. Tumor resection was performed without any other treatments such as chemoradiotherapy. No recurrence has so far been detected for three years.

Discussion

FDCS is a very rare tumor believed to originate in FDC and was first reported in 1986 by Mondo et al [2]. Majority of the cases show the presence of such tumors in the lymph nodes, with reports of approximately 80 previous cases occurring in the head and neck region, with a predilection for the tonsils [3-5]. FDCS has been reported in a wide range of patient ages and equally in both sexes.

The etiology of the disease is unknown but may be associated with hepatitis B virus infection or Cattleman's disease [6]. This patient tested negative for Epstein-Barr virus-encoded small RNAs by *in situ* hybridization. Patients are usually negative for CD79a, but our patient was positive for it; in addition, some patients are reported to be negative for UCHL1 (CD45RO) but some other are reported to be positive. Thus, FDCS can have a variety of immunological characteristics. In one report, 26 of 46 cases of FDCS in the head and neck region were initially misdiagnosed; accurate diagnoses were reached only after reassessment. Immunohistochemical staining has shown positive staining for FDC markers including CD21, CD23, CD35, Ki-M4p, CAN.42, and D2-40 [7].

FDCS is a low-grade to intermediate-grade tumor [6] with distant metastasis in 16% of cases originating in the head and neck region and tumor-specific death in 10%. The published disease-free survival rate is 66.2% [8]. In tumor grade classification for estimating prognosis, Ki-67 labeling index <10% is considered low grade and ≥10% is considered as high grade [8]. Treatment varies and includes surgery alone, combination therapy with surgery and radiotherapy or chemotherapy, or chemoradiotherapy [9]; in addition, no consensus has been reached regarding the preferred treatment. Patients with recurrence or tumor progression have been treated with cyclophosphamide, doxorubicin, vincristine, and prednisone, which are used in treating

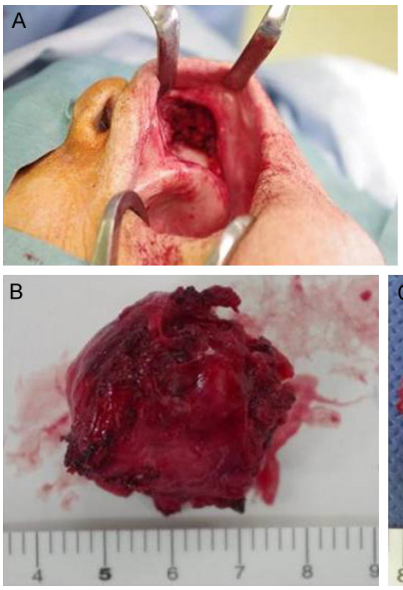

Figure 2. Macroscopic findings: A. Surgical appearance. B. The surface of the excised tissue was smooth. C. The cross-sectional surface of the tumor was grayish white.

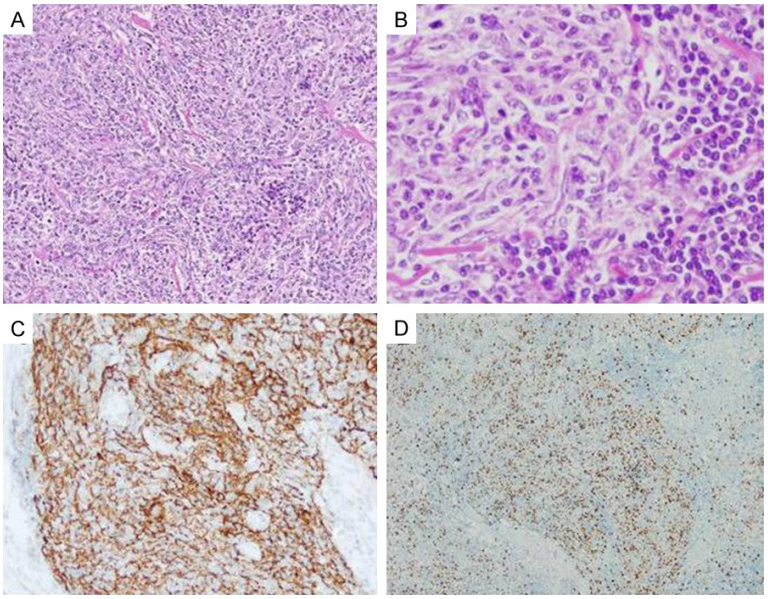

Figure 3. Microscopic findings: A (original magnification, ×100) and B (×400): Whorl formations of tumor cells with unclear cell borders and growing as a sheet. C: CD21-positive immunostaining of the tumor cell cytoplasm (×100). D (×100): MIB-1 index was 70%, exclusive of non-tumor cells, in hot spots (×100).

lymphoma [10-12]. In this patient, the MIB-1 index was 70%; hence, the tumor was believed to be high grade; however, given the advanced age of the patient and considering her overall health condition, additional treatment such as radiotherapy or chemotherapy was not provided. To our knowledge, this is the first report of FDCS occurring in the cheek. Because it is such a rare tumor, it is necessary to compile case reports from around the world to establish a uniform treatment strategy.

Conclusion

To our knowledge, this unusual case is probably the first reported one of subcutaneous FDCS occurring in the cheek. Given that the etiology and preferred treatment are currently not established, additional experiences are needed before recommendations can be made.

Disclosure of conflict of interest

None.

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