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Case Report

Atypical Axillary Mass: Follicular Dendritic Cell Sarcoma within Castleman's Disease: A Case Report and Review of Literature

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Abstract

Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm (0.4% of soft tissue sarcomas) originating from follicular dendritic cells. On the other hand, Castleman's disease (CD) is a rare lymphoproliferative disorder first described in 1956 by Castleman et al. as a benign, localized enlargement of hyperplastic lymph nodes. Castleman's disease has been found in association with FDCS in a minority of cases, suggesting that it may represent a precursor lesion. Combination of both FDCS and unicentric CD is extremely rare.

As far as we know, there are no documented cases of a follicular dendritic cell sarcoma within a hyaline-vascular Castleman's disease localized in the axilla.

There is no consensus about treatment strategy in this situation; surgical resection when possible seems to be the best option, with or without complementary radiotherapy.

We present the case of a 42-year-old male with a single axillary mass identified as a FDCS in a unicentric CD in the axilla.

Introduction

Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm (0.4% of soft tissue sarcomas) originating from follicular dendritic cells; they usually present as coalescent nodal mass of variable size. Due to its rarity, most of the data on FDCS are based on case reports or small case series.

Castleman´s disease has been found in association with FDCS in a minority of cases, suggesting that it may represent a precursor lesion [1]. FDCS generally affect young to middle-aged adults, with a mean age of 43 years with no sex predilection and they usually present as painless coalescent nodal mass (60%) of variable size (1-20cm diameter). The optimal treatment for FDCS is yet to be found due to the limited experience but it has a slight recurrent and metastatic potential.

Castleman's disease (CD) is a rare lymphoproliferative dis-

order first described in 1956 by Castleman et al as a benign, localized enlargement of hyperplastic lymph nodes. It can occur at any age with a peak incidence in the third and fourth decades [2]. It is divided into two groups depending on its histologic and clinical features. In 1972, Keller et al sub classified CD into a hyaline-vascular type an a plasma-cell type based on their histologic features. Some patients have a mixed form. Clinically, CD can be divided into a localized form, which refers to a unicentric (UC) disease, and a widespread disease (multicentric) (MC).

Combination of both FDCS and UC CD is extremely rare. We present the case of a 42-year-old male with a FDCS and a unicentric CD in the axilla.

Case Report

A 42-year-old male presented with a single axillary mass developed 15 years ago that had recently grown. He had no other past medical history.

Physical examination showed a solid axillary mass with no peripheral nerve involvement symptoms, neither vascular complications. The laboratory tests were normal. A thoraco-abdomino-pelvic computed tomography (CT) scan demonstrated a right axillary mass of 4.7 x 5.3 cm, with heterogeneous contrast captation, suggesting a pathologic adenopathy. There were no other pathologic findings.

We performed a complete radical surgical resection and there were no postoperative complications.

Macroscopically, the resected piece weight 240gms and sized 12x7x6cms. The consistence was slightly firm. (Figures 1,2).

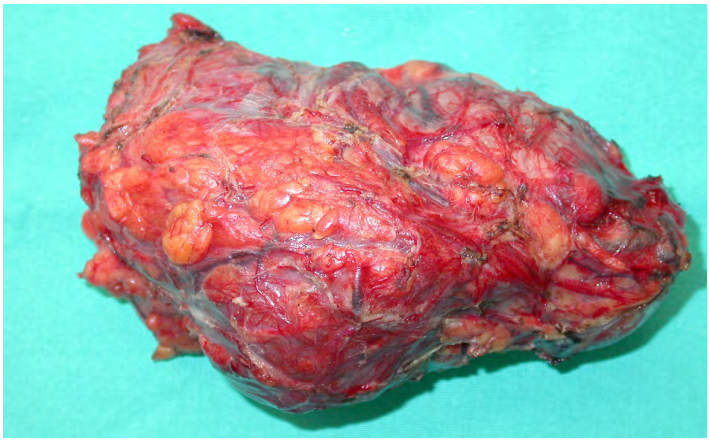


Figure 1

Microscopically the tumor was composed of fusiform cells arranging in short fascicular pattern inside a lymph node that conserved the follicular architecture (figures 3,4, 5 and 6). There were areas with capillary vascular hyalinization and peripheral disposal of B lymphocytes, with a row pattern. Another characteristic feature was the sprinkling across the entire tumor of B and T lymphocytes. Immunochemical analysis was positive for CD21 and CD23, and slightly for D2-40. In the tumoral area CD23 was negative. CD 68 and S100 were positive for non tumoral dispersed cells.

Patient is asymptomatic and disease free.

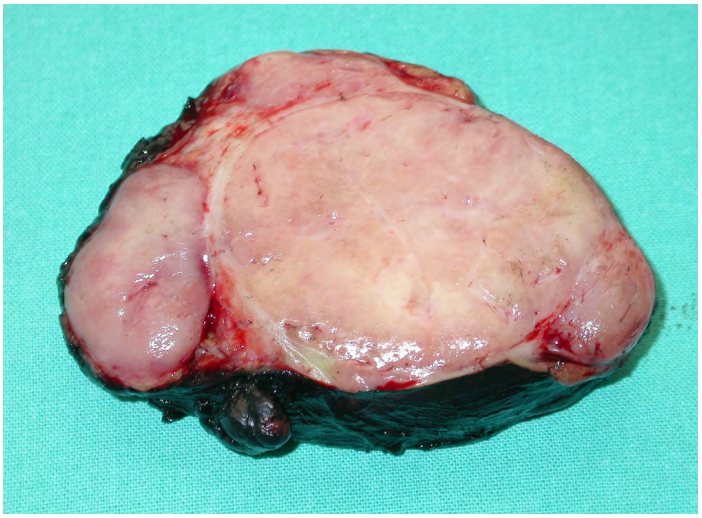
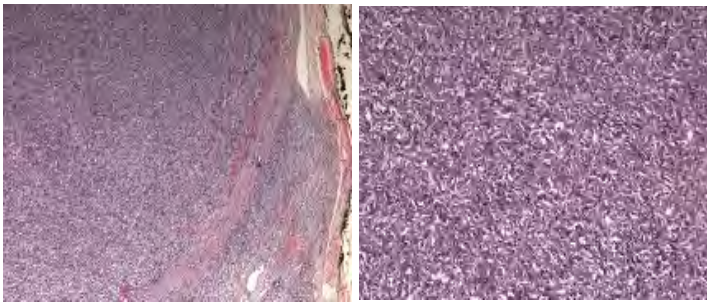


Figure 2

Figures 1-2. Surgical *en bloc* resection.



Figures 3-4. (H&E staining, x400).

Figures 5-6. Hyalinization and low mitotic index.

Discussion

Follicular dendritic cell sarcoma is a rare neoplasm (0.4% of soft tissue sarcomas) originating from follicular dendritic cells; these cells are non-lymphoid, non-phagocytic accessory cells of the lymphoid system and play an integral role in regulation of the germinal center reaction and presenting antigens to B-cells [3]. Due to its rarity, most of the data on FDCS are based on case reports or small case series.

Follicular dendritic cell sarcoma (FDCS) arises from follicular dendritic cells localized within lymphoid follicles. There is no definite etiology for most cases of FDCS, but interestingly, Castleman´s disease has been found in association with FDCS in a minority of cases, suggesting that it may represent a precursor lesion [1]. Haiwei et al suggest that FDCS proliferation and dysplastic changes occurring in Castleman´s disease can form the background from which a FDCS develops. A possible role for p53 has also been proposed, with overexpression of p53 protein noted in FDCS as well as an increased number of weakly p53-positive spindle-cells in a hyaline-vascular Castleman's disease specimen [1]. Other studies associate FDCS with Epstein Barr virus (12%) because FDCS expresses CD21 (EBV receptor), especially those occurring in the liver and spleen [3].

It generally affects young to middle-aged adults, with a mean age of 43 years with no sex predilection. They usually present as painless coalescent nodal mass (60%) of variable size (1-20cm diameter), but extranodal disease is also commonly seen in the head and neck (tonsil, nasopharynx) and intraabdominal organs (in particular the pancreas and peripancreatic tissues). Other locations are as well: axilla, gastrointestinal tract (17%), liver, oral cavity (13%), mediastinum (8%) and spleen (2%) [3].

The pathologic characteristics reveal oval to spindle cells with elongated nuclei, dispersed chromatin and pale eosinophilic cytoplasm. Lymphocytes may appear around blood vessels with a cuffing pattern or as a concentric whorl. The most widely used markers are CD21 and CD35. Other useful markers are vimentin, CD23, CD68, S-100 protein, fascin, Ki-M4p and Ki-FDC1p; however, these are unspecific.

The optimal treatment for FDCS is yet to be found due to the limited experience but some studies suggest that surgical radical resection avoids need for radiotherapy or chemotherapy. In fact, Saygin et al [4], suggest that surgery should be the mainstay treatment for early FDCS cases, as the OS is better in patients treated with surgery than other treatment modalities. In any case, FDCS has a slight recurrent and metastatic potential and it should be viewed as a low-grade malignancy [1], but literature reports show a wide treatment variety, including adjuvant chemotherapy and radiotherapy. Further investigations are required to convincingly demonstrate optimal treatment strategy, although surgical resection is the preferred first option [5].

Castleman's disease (CD) is a rare lymphoproliferative disorder first described in 1956 by Castleman et al as a benign, localized enlargement of hyperplastic lymph nodes. It can occur at any age with a peak incidence in the third and fourth decades [2]. It is divided into two groups depending on its histologic and clinical features. In 1972, Keller et al subclassified CD into a hyaline-vascular type a plasma-cell type based on their histologic features. Some patients have a mixed form. Clinically, CD can be divided into a localized form, which refers to a unicentric (UC) disease, and a widespread disease (multicentric) (MC) [6]. CD occurs in the thorax in 70% of cases, abdomen and pelvis in 15%, and in the neck in 10-15% of cases [2]. UC CD is predominantly developed in the mediastinum or the lung hilum. However, the abdomen and the retroperitoneum may also be involved [5].

The etiology is unknown, but it has been described in association with human immunodeficiency virus infection (HIV), human herpesvirus 8 infection (HHV-8), Kaposi´s sarcoma, Hodgkin´s lymphoma, non-Hodgkin´s lymphoma, and POEMS syndrome (poly neuropathy, organomegaly, endocrinopathy, M protein and skin changes) [2,8].

The clinical presentation of the unicentric type of Castleman´s disease is associated with mass-effect symptoms related to the compression of adjacent organs. Patients also complain of unspecific symptoms such as postprandial discomfort, vomiting, weight loss, urinary retention and abdominal or lumbar pain, which makes clinical diagnosis very difficult. Thus, other evaluations and complementary studies are needed to identify Castleman´s disease. CT scan may reveal a hypervascularized mass with heterogeneity, suggesting a lymphoma, soft-tissue sarcoma or vascular tumors.

As explained before, CD is histopathologically subclassified into two types. The HV type is characterized by giant lymph follicles centered on central vessels with marked hyalinization. The PC type contains many more polyclonal plasma cells with a less marked hyalinization and vascularization. Approximately 90% of all localized forms are HV type, but no differences in outcomes have been found comparing the PC and the HV types in patients with UC or MC disease. However, UC type is superior to MC type in terms of clinical outcomes and long-term follow-up results.

It is very important to determine if it is UC or MC, because treatment becomes different; while in MC form the role of surgery is limited , in UC form, the *en bloc* resection of the tumor is the best treatment option, with no further treatment needed when resection is successful [7].

Combination of both FDCS and UC CD is extremely rare. As

far as we know, there are no documented cases of a follicular dendritic cell sarcoma within a hyaline-vascular Castleman´s disease in the axilla. There is no consensus about treatment strategy in this situation; surgical resection when possible seems to be the best option, with or without complementary radiotherapy [9]. We performed an *en bloc* resection and complementary local radiotherapy. Patient is asymptomatic and disease free, but long-term results are not available yet.

Conclusion

Follicular dendritic cell sarcoma is a rare neoplasm (0.4% of soft tissue sarcomas) originating from follicular dendritic cells; due to its rarity, most of the data on FDCS are based on case reports or small case series. It generally affects young to middle-aged adults, with a mean age of 43 years with no sex predilection.

Castleman's disease (CD) is a rare lymphoproliferative disorder first described in 1956 by Castleman et al as a benign, localized enlargement of hyperplastic lymph nodes. CD can be divided into a localized form, which refers to a unicentric (UC) disease, and a widespread disease (multicentric) (MC).

Some authors suggest that FDCS proliferation and dysplastic changes occurring in Castleman´s disease can form the background from which a FDCS develops, but there is few evidence of it due to lack of cases.

Combination of both FDCS and UC CD is extremely rare. We have not found documented cases in literature of a follicular dendritic cell sarcoma within a hyaline-vascular Castleman´s disease in the axilla.

There is no consensus about treatment strategy in this situation, but surgical resection when possible seems to be the best option, with or without complementary radiotherapy. More studies need to be performed to establish the relation between FDCS and CD and to define a treatment strategy. Although it is considered to have a slight metastatic potential, prognosis is unclear.

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