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Giant Follicular Dendritic Cell Sarcoma of Cervical Lymph Node: A Pediatric Case Report

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ABSTRACT

Follicular dendritic cell sarcoma of the neck (FDCS) is a rare neoplasm with only 7 reported cases for pediatric patient.

A 13 year-old girl presented with a left neck tumor. Positron Emission tomography coupled with a cervical scan showed a large left cervical lymphadenopathy hypermetabolic superior jugulocarotid.

Fine-needle aspiration cytology are performed which confirmed a diagnosis of FDCS.

The definitive diagnosis was reached by the Immunohistochemical study that showed tumor cells expressing CD23+, CXCL13+, and CD68+. Some of them are experimenting with CD21+, others cells, rarer, experimenting with S100.

The therapeutic decision was a left cervicotomy associated excision of the mass with a homolateral cervical dissection.

INTRODUCTION

Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm with only 7 reported cases for pediatric patient [1-3].

Follicular dendritic cells (FDCS) are nonlymphoid cells essential for antigen presentation presenting in B lymphofollicles. They are spindle-shaped multinucleate cells with dendritic projections. Some cells can be binucleated or multinucleated. Benign or malignant tumors originating from FDCs rare and only a limited number of cases have been reported [4,5].

The diagnosis of FDCS is made by its typical location in a lymph node, morphology, immunohistochemical findings, and in some cases, ultrastructural findings. We report a case of FDCs. The clinicopathological features and the diagnosis and treatment of this rare tumor is discussed.

AIM

Illustrate a clinical case for pediatric patient of Follicular dendritic cell sarcoma of the cervical lymph node by emphasizing the contribution of the fine need aspiration in the diagnosis and to discuss the different therapeutic means of this affection.

CASE REPORT

A 13-year-old girl, without significant pathological history, presented at the consultation with a left cervical mass without dysphagia, dyspnea or dysphonis.

The mass gradually increases in size over 4 months. Physical examination found a hard mass of 4cm, and the skin opposite was normal.

The blood count with HIV, toxoplasmosis and Epstein Barr virus serology was negative.

A cervical scanner showed a homogeneous upper left oval jugulocarotid lymphadenopathy without central necrosis measured 35mm in diameter **(Figure 1).**

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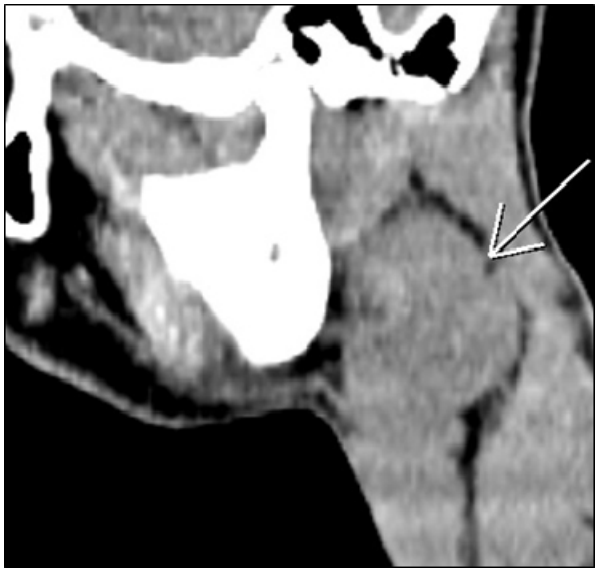

Figure 1. Sagittal cervical tomography showing a lymphadenopathy in the upper left jugular region**.**

Positron emission tomography coupled with a Cervical scan showed a large left cervical lymphadenopathy hypermetabolic superior jugulocarotid SUVmax = 7.2 measured 35mm without other secondary localizations **(Figure 2).**

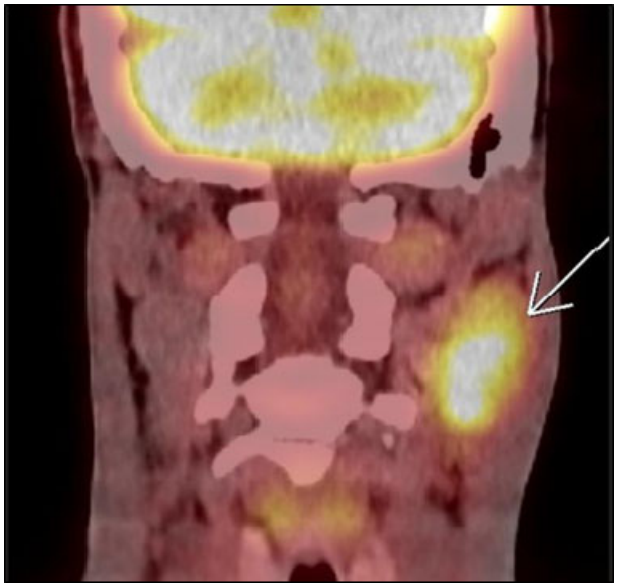

Figure 2. Large left cervical lymphadenopathy hypermetabolic superior jugulocarotid SUVmax = 7.2.

Fine needle aspiration cytology was performed which showed destruction of the normal ganglion near architecture by spindle cells in a stroma rich in lymphocytes and including some B immunoblasts **(Figure 3).**

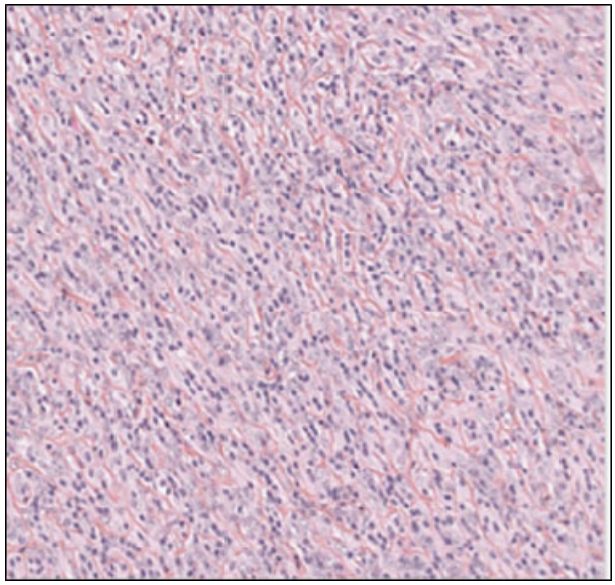

Figure 3. Photomicrograph showing a diffuse proliferation replacing normal lymph node architecture.

These spindle cells sometimes have a bilobed nucleus of the dendritic follicle cell type with a large central nucleolus **(Figure 4).**

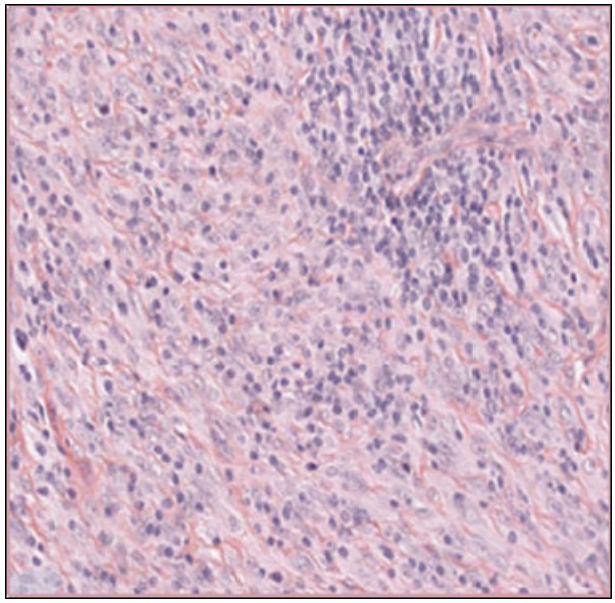

Figure 4. Spindle cells with a bilobed nucleus of the dendritic follicle cell type with a large nucleolus.

These cells are organized in a basic manner and the phenotype CD3 +, CD21 +, CXCL13 +, D2-40 +, CD23+ **(Figure 5).**

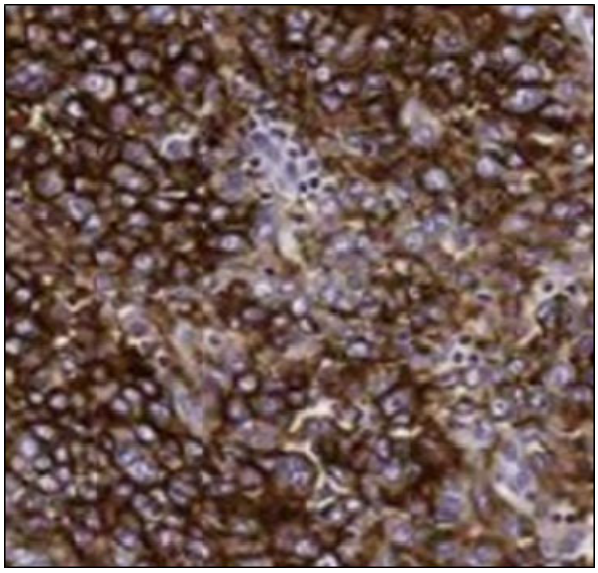

Figure 5. Neoplasic cells express CD 23.

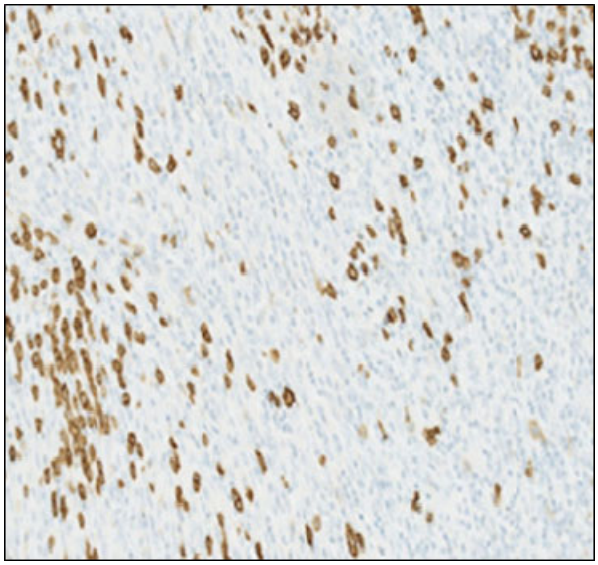

Figure 6. Neoplasic cells express CD20.

Immunostaining with anti-CD20 finds some residual follicles.

EBV search is negative.

The therapeutic decision was a left cervicotomy associated excision of the mass with a homolateral cervical dissection.

The histological of the excised mass and the lymph nodes showed spindle-shaped or ovoid tumor cells, their cytoplasm moderately eosinophilic, their nucleus is rounded rather monotonous with fine chromatin and a visible central nucleolus and absence of ganglionic tumor proliferation.

An immunohistochemical study showed tumor cells expressing CD23+, CXCL13+, and CD68+. Some of them are experimenting with CD21+, other cells, rarer, experimenting with S100.

CD3 and CD20 are positive on entangled cells and negative on tumor cells **(Figure 6)** the diagnosis of FDCS was confirmed.

DISCUSSION

FDSC is a rare neoplasm for patient pediatric with only 7 cases found in the literature. The first case was describe by Apostolos [1].

FDC sarcoma is a very rare neoplasm which was first characterized by Monda [6].

A review of the literature suggests that FDCS has a slight male predominance, and the median age of diagnosis is 40 (age 9-86) [1-5].

The authors discuss the use of fine-needle aspiration for the pre-surgical diagnosis of FDC sarcomas and eliminate other differential diagnoses such as paraganglioma or undifferentiated carcinoma that can be help on the management of this neoplasm [7,8].

Several radiological examinations are requested in case of a cervical mass such a resonance magnetic imaging or positron emission computed tomography but remain non-specific in case of follicular dendritic cell sarcoma.

Histologically, the tumor cells are spindly, ovoid, or polygonal in shape, have eosinophilic cytoplasm, and have indistinct borders, resulting in a syncytial appearance [5,9].

They contain oval to round nuclei with smooth borders and mild atypia.

Immunohistochemically the most sensitive and specific markers for FDCS are CD21, CD23, and CD35 [5,10].

Variably markers found in FDCS include S-100, and vimentin, whereas nonspecific include desmoplakin [4,11].

Phenotype CD68 are nonspecific markers for FDCS [12].

The chemokine CXCL13 and podoplanin (D2-40) produced by the neoplastic FDCS cells can be used as a biomarker to diagnose this tumor [13,14].

An immunohistochemical study on our fine need aspiration showed the positivity of phenotype CD21, CD23, CXCL13 and D2-40.

However, following markers CD23, CD68 and on rare cells, s100 were positive on the removed mass.

The management of FDCS is similar to that of other soft tissue sarcomas; surgical resection remains the standard treatment.

The role of adjuvant treatment in the management of FDCS remains uncertain.

Some authors have suggested adjuvant radiotherapy, and others have recommended chemotherapy or radiotherapy only when the tumor is aggressive, of high volume, and surgically unresectable [9,15,16].

A recent series compared patients who underwent surgical resection alone with those who underwent surgical resection with adjuvant radiotherapy. Their results showed that radiation therapy reduced the rate of recurrence, but much larger studies are needed to confirm this [17].

In the case that we have reported, the decision of the multidisciplinary meeting between surgeons, oncologists and radiotherapists was taken not to add radiotherapy, avoiding complications of radiation therapy such as radio necrosis of the mandible and neuropathy, to the treatment with close monitoring because the lymphadenopathy is completely removed with no capsular breakage and absence of positive lymph nodes recess performed.

CONCLUSION

Follicular dendritic cell sarcoma (FDCS) of the lymph node is a rare neoplasm for the patient pediatric that can be diagnosed on the fine needle aspiration.

The management of FDCS is similar to that of other soft tissue sarcomas, surgical resection remains the standard treatment.

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