CLINICAL CASE

Follicular dendritic cell sarcoma. Case report

Sarcoma de células dendríticas foliculares. Informe de un caso

Antonio S. Sosa-Luis[1], Claudia T. Gutiérrez-Quiroz[1], Francisco Espinosa- Zúñiga[1], Daniel E. Meléndez-Mena[1], Álvaro J. Montiel-Jarquín[1] and Jorge Loría-Castellanos[2]*

[1]High Specialty Medical Unit, Puebla Specialty Hospital, Instituto Mexicano del Seguro Social (IMSS); [2]Division of Special Health Projects, IMSS. Puebla, Puebla, Mexico

Abstract

Background: *Follicular dendritic cell sarcoma is a rare pathology, it occurs mainly in nodal sites such as head, neck, oropharynx, although extranodal presentation such as spleen and liver may occur. In most cases it is asymptomatic but may present general symptoms, abdominal pain or fever. Immunohistochemistry is essential to make a definitive diagnosis.* **Case presentation:** *Forty-year-old woman, with submaxillary lesion, in the neck, right fronto-parieto-occipital region with dry oral mucosa. It was initially managed as a Sjögren's syndrome ruled out by the histopathological result of salivary gland biopsy. Subsequently, a neck ganglion biopsy was performed that reported follicular dendritic cell sarcoma, with positive immunohistochemical expression for CD23 and negative for CD21 and LCA. It was managed with samarium with a survival of 3 months from the time of its diagnosis.* **Conclusions:** *Follicular dendritic cell sarcoma is rare and its global survival is short.*

Keywords: *Sarcoma. Dendritic cells. Immunohistochemistry. Case report.*

Resumen

Antecedentes: *El sarcoma de células dendríticas foliculares es una enfermedad rara, caracterizada principalmente en sitios nodales como la cabeza, el cuello y la orofaringe, aunque puede ser extranodal, como en el bazo y el hígado. En la mayoría de los casos cursa asintomática, pero puede presentar síntomas generales, dolor abdominal o fiebre. La inmunohistoquímica es indispensable para llegar a un diagnóstico definitivo.* **Presentación del caso:** *Mujer de 40 años, con abultamiento submaxilar en el cuello, región frontoparietooccipital derecha, y sequedad de mucosa oral. Se manejó inicialmente como un síndrome de Sjögren, que fue descartado por el resultado histopatológico de la biopsia de glándula salival. Posteriormente se realizó biopsia de ganglio del cuello, que reportó sarcoma de células dendríticas foliculares con expresión inmunohistoquímica positiva para CD23 y negativa para CD21 y ACL. Se manejó con samario y tuvo una sobrevida de 3 meses desde el diagnóstico.* **Conclusiones:** *el sarcoma de células dendríticas foliculares es raro y la sobrevida es corta.*

Palabras clave: *Sarcoma. Células dendríticas. Inmunohistoquímica. Reporte de caso.*

***Correspondence:**
Álvaro J. Montiel-Jarquín
2 norte 2004
Col. Centro
C.P. 72000, Puebla, Pue., México
E-mail: dralmoja@hotmail.com;
Alvaro.montielj@imss.gob.mx

Date of reception: 10-02-2020
Date of acceptance: 20-01-2021
DOI: 10.24875/CIRUE.M23000356

Cir Cir (Eng). 2021;89(4):525-528
Contents available at PubMed
www.cirugiaycirujanos.com

Introduction

Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm characterized by follicular dendritic cells differentiation. It was described by Monda et al. in 1986[1] and, so far, about 400 cases have been reported in the world literature[2]. It can occur in the neck, the mediastinum, the spleen, the liver, and other sites, such as the oropharynx.

Symptoms depend on tumor location; when it occurs in the neck, it can be asymptomatic, although some patients experience abdominal pain or systemic manifestations such as fever, fatigue and nocturnal diaphoresis[2,3,4,5]. Evolution is variable. Currently, there is no well-established treatment strategy, and its prognosis is poor for life in the short term. The purpose of this study is to present the clinical case of a woman with FDCS in the neck, her evolution and her treatment.

Clinical case

Forty-year-old woman in whom her ailment had started 6 months earlier, when she noticed a submaxillary bulge, in the neck right fronto-parieto-occipital region, with oral mucosa dryness. It was managed as a case of Sjögren's syndrome, but salivary gland biopsy histopathological report showed no diagnostic criteria for it. Subsequently, the patient underwent diagnostic workup, with blood count report with depletion of all three series: hemoglobin 11.6, platelets 19,000, white blood cells 1,300, creatinine (2.85 mg/dL) and lactate dehydrogenase elevation (713 U/L); coagulation and liver function tests were normal. Head and neck computed tomography reported a lateral neck mass of origin to be determined, without ruling out a neoplastic process being possible; adenopathy at level II on left side of the neck and at levels II, III, and IV on right side (Fig. 1). Bone scan showed bilateral fronto-parietal, sacroiliac, femoral and ischial lytic lesions (Fig. 2).

Owing to the blood count alteration and advanced clinical stage, the patient was not considered to be candidate for surgery, which is why she only underwent biopsy of the neck bulge area.

Histopathological report indicated FDCS (Fig. 3), and immunohistochemistry showed negative expression for CD21, positive for CD23 and negative for leukocyte common antigen (LCA) (Figs. 4 to 6).

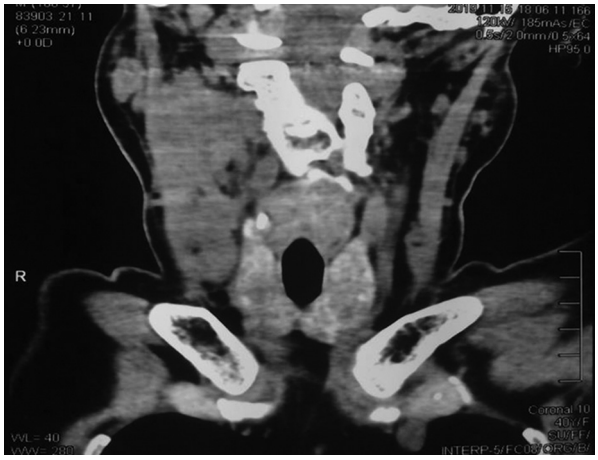

Figure 1. *Computed tomography: lateral lesion of the neck of origin to be determined, adenopathy at level II of left side of the neck and at right-side levels II, III and IV.*

Treatment with samarium at a dose of 3.4 mR/h was established. The patient had radiotherapy sessions scheduled, which were not concluded because she was in poor general condition, with fatigue, asthenia and adynamia, which required hospital admission. During this hospitalization, anemia, thrombocytopenia and leukocytopenia were documented. She had blood products transfused, and died 8 days later. Survival of this patient was 3 months from diagnosis to death.

Discussion

FDCS is a neoplastic proliferation that implies the fusion of ovoid cells with morphology and phenotypic characteristics similar to those of normal follicular dendritic cells. The World Health Organization classifies it as a histiocytic and dendritic cell neoplasm[3]. Dendritic cells are non-myeloid, non-phagocytic cells of innate immunity, which are found primarily in germinal centers and secondarily in lymphoid organs, and are characterized for expressing CD21, CD35, R4/23 and KiM4e on their surface[3]. FDCS can occur at lymph nodes and extranodal sites, such as the oral cavity, nasopharynx, gastrointestinal tract, liver, pancreas, and spleen[5]. Pang et al.[2], in a MEDLINE review, published two cases of FDCS in men and reported 97 additional cases over a 46-year period (1978 to 2014), with 40% of cases occurring in cervical lymph nodes, 24% in the oropharynx, and 10% in soft tissues of the neck. The youngest ages (30.1 ± 16.5 years) at presentation were in patients with soft tissue lesions of the neck, and the oldest ages (61.5 ± 17.0 years), in subjects with thyroid neoplasms with

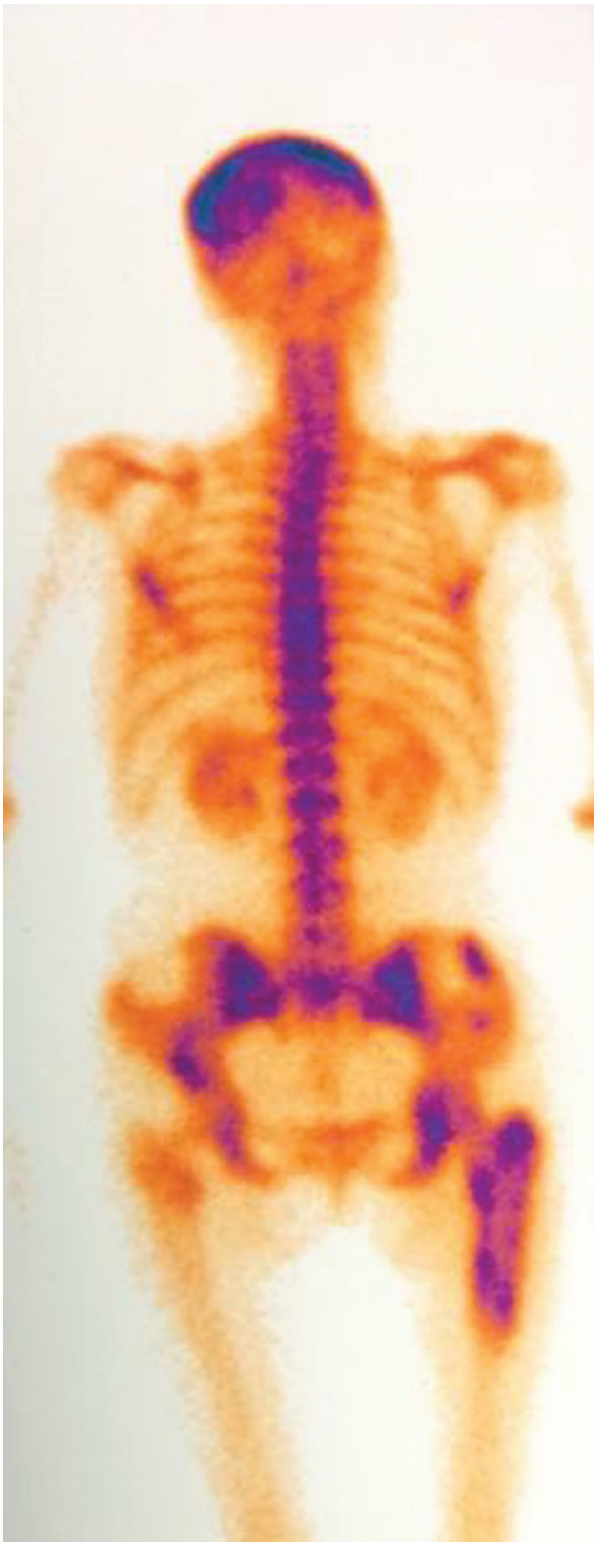

Figure 2. *Bone scan: osteoblastic activity increase is observed at frontal and parietal bones, sacroiliac joints, bilateral ischium and femur.*

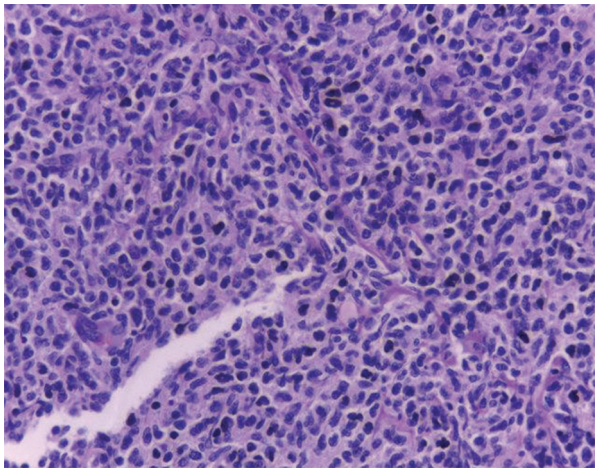

Figure 3. *Hematoxylin & eosin staining (x 40): cells with an histiocytic appearance are observed, with apparent nucleolus and some of them with a slit.*

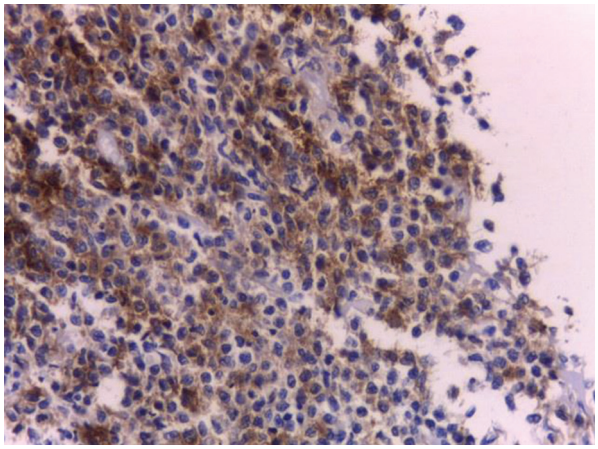

Figure 4. *Immunohistochemistry: CD21 negative expression.*

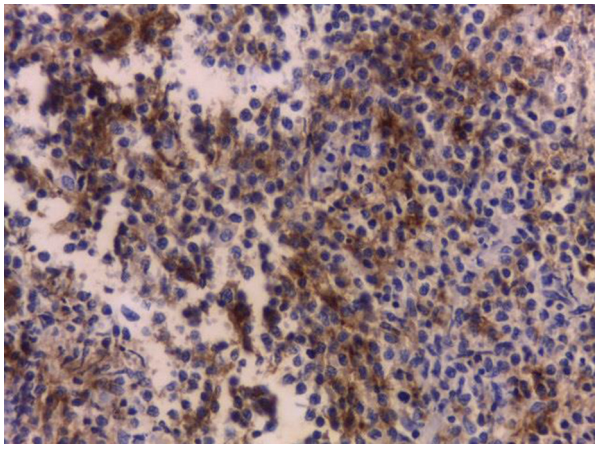

Figure 5. *Immunohistochemistry: CD23 positive expression.*

a slight predominance of occurrence in men (53%), and mean tumor diameter of 4.5 cm. Another study reports 14 cases of FDCS over the 1995 to 2005 period, at M.D. Anderson Cancer Center, out of which 21.42% had cervical lymphadenopathy; 35.71%, abdominal lymphadenopathy; 21.42%, mediastinal tumors; 14.28%, nasopharyngeal disease; and 7.14%,

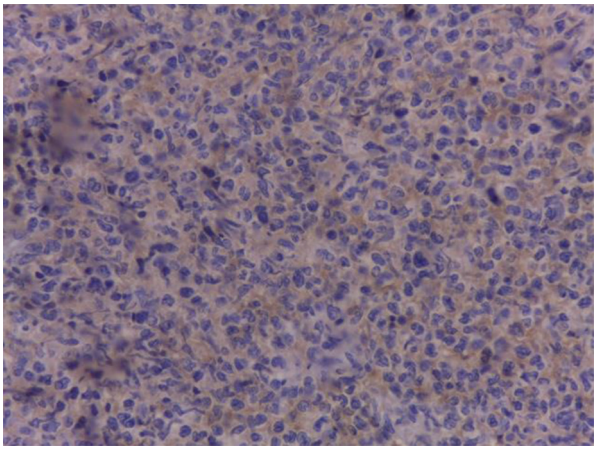

Figure 6. *Immunohistochemistry: LCA negative expression.*

pleural effusion[6]. Patients are usually asymptomatic and the only clinical sign may be a painless, slowly-growing cervical lymphadenopathy[3]. These neoplasms are related to paraneoplastic syndromes, such as pemphigus, myasthenia gravis and Castleman disease[2,3]. There are immunohistochemistry markers that support the diagnosis, but they are non-specific. Some, such as CD21, CD23 and CD35, are frequently expressed in FDCS, and there are other markers, such as CD68, vimentin and S100, which may be expressed, but are more common in pathologies such as undifferentiated pleomorphic sarcomas, Langerhans cell histiocytosis and schwannomas, respectively[1,6].

Differential diagnoses for FDCS include interdigitating dendritic cell sarcoma, thymomas, spindle cell carcinomas, undifferentiated carcinoma metastases, malignant melanoma, and gastrointestinal stromal cell tumors (GIST)[3].

Surgical resection can be carried out when clinical stage and patient conditions allow it. Combined treatment with surgery, radiotherapy and chemotherapy appears to be the best option, with an overall survival of up to 8 years[6,7].

FDCS has a variable clinical course, and there is currently no standard treatment strategy. Its prognosis is poor, with a short survival, which depends on the clinical stage the disease is detected at. Local recurrence is common (40-50% of reported cases) and metastases from lymph node tumors occur in approximately 10% of patients, in comparison with extranodal tumors, which have a higher metastatic potential (20%)[2,3].

It is important to highlight the usefulness of immunohistochemistry techniques in the differential study of this type of lesions, due to the high sensitivity and specificity of CD21, CD35 and CD23 antibodies, as demonstrated by Jorge-Buys et al.7 in their five-case series.

The presented case, diagnosed at clinical stage IV, exhibited high morbidity and a short survival (3 months), which is consistent with the world literature.

Funding

This research did not require external funding.

Conflicts of interest

The authors declare that they have no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures that were followed adhered to the ethical standards of the responsible committee for experimentation on human subjects and were in agreement with the World Medical Association and the Declaration of Helsinki.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained informed consent from the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

References

1. Monda L, Warnke R, Rosai J. A primary lymph node malignancy with features suggestive of dendritic reticulum cell differentiation. A report of 4 cases. Am J Pathol. 1986;122:562-72.
2. Chen T, Gopal P. Follicular dendritic cell sarcoma. Arch Pathol Lab Med. 2017;141:596-9.
3. Pang J, Mydlarz WK, Gooi Z, Waters KM, Bishop J, Sciubba JJ, et al. Follicular dendritic cell sarcoma of the head and neck: case report, literature review, and pooled analysis of 97 cases. Head Neck. 2016;38(Suppl 1):E2241-9.
4. Wu A, Pullarkat S. Follicular dendritic cell sarcoma. Arch Pathol Lab Med. 2016;140:186-90.
5. An P, Li Y, Zhao J. Follicular dendritic cell sarcoma of the parotid gland: a case report. Oral Oncol. 2020;102:104515.
6. Soriano AO, Thompson MA, Admirand JH, Fayad LE, Rodriguez AM, Romaguera JE, et al. Follicular dendritic cell sarcoma: a report of 14 cases and a review of the literature. Am J Hematol. 2007;82:725-8.
7. Jorge-Buys DL, Lastra-Camacho G, Campos-Martínez J, Romero-Guadarrama M, Ortiz-Hidalgo C. Sarcoma de células dendríticas foliculares de localización ganglionar y extraganglionar. Estudio clinicopatológico e inmunohistoquímico de cinco casos. Cir Cir. 2008;76:145-52.