Follicular dendritic cell tumour of the cervical lymph node: case report and brief review of literature

Case report: il tumore/sarcoma a cellule follicolari dendritiche

U. MOZ, U. PIGNATELLI, P. FORNER, E. CANDIAGO, G. D’ADDAZIO, C. CASIRATI, P. GAMBA
Department of Otorhinolaryngology; 1 Department of Histopathology, Poliambulanza Hospital, Brescia, Italy

Summary

Follicular dendritic cell tumour/sarcoma is a rare tumour involving lymph-node or extra-lymph-node sites; review of the literature reveals very few cases of follicular dendritic cell tumour, probably since, in the past, the disease has often been mistaken for other neoplasms: low differentiated carcinomas and fusate cell carcinomas, sarcomas, melanomas, thymic neoplasms, Castle carcinoma and other dendritic cell tumours (especially interdigital cell tumour/sarcoma). In the case described here, attention is focused on the diagnostic difficulties and on the therapeutic profile, comparing data with those reported in the international literature.

Riassunto

Il tumore/sarcoma a cellule follicolari dendritiche è una rara neoplasia che può colpire sia il distretto linfonodale sia quel-lo extralinfonodale; dall’analisi della letteratura emergono discordanze principalmente legate alla difficoltà, specie in passato, di riconoscere questa malattia e di differenziarla da altre: carcinomi scarsamente differenziati e a cellule fusate, sarcomi, melanomi, neoplasie ad origine timica, carcinoma con aspetti di differenziazione timica (Castle) altri tumori ad origine dalle cellule dendritiche (in particolare il tumore/sarcoma a cellule interdigitate). La discussione di un caso clinico offre lo spunto per evidenziare il comportamento clinico e gli aspetti terapeutici e per un confronto con i più recenti dati emersi in letteratura.

Introduction

Follicular dendritic cell sarcoma is a rare neoplasm that interests both sexes and mainly the adult age; this neoplasm is associated with Castleman’s disease in about 10-20% of cases: Castleman’s disease can occur prior to the onset of follicular dendritic cell tumour.

Follicular dendritic cell tumour site may be:
- Lymph nodes, particularly cervical, mediastinal, axillary, intestinal or retro-peritoneal;
- Extra-lymph nodes, for example, tonsil, oral cavity, spleen, intestine, liver, soft tissue or skin.

This tumour can relapse or metastasise, even many years later, particularly on lymph nodes, lungs, liver and breast.

Differential diagnosis is given by: low differentiated carcinomas and fusate cell carcinomas, sarcomas, melanomas, thymic neoplasms, Castle carcinoma and other dendritic cell tumours (especially interdigital cell tumour/sarcoma).

The aim of the present research is to demonstrate that immunohistochemical markers CD21, CD23, CD35, Vimentin and EMA are very important for differential diagnosis (Figs. 1-4).

This report deals with a case of painless swelling of the neck, focusing attention on the difficulties encountered in the differential diagnosis. The most appropriate instrumental investigations and best therapeutic approach are discussed.

Case report

A 56-year-old white female non-smoker, with a past medical history of surgery for breast cyst and follicular right-sided thyroid adenoma and a present medical history of hypertension.

The patient came to our attention with a left-sided level II swelling of the neck, without fever, which she had first noticed 30 days previously. During ENT evaluation, we found one 3 cm hard-elastic painless lymph node (levels IIA-IIB) with partial mobility. Ultrasonography (US) of the neck showed evidence of one 2.5 cm left-sided lymph node at level II. Fine needle aspiration biopsy (FNAB) was positive for malignant cells without possibility of discrimination; the immunohistochemical pattern was not typi-
cal of a definite tumour but rather of an epithelial tumour with high grade malignancy.
In the present case, we obtained incorrect results from FNAB and relative cytology, on account of:
- CAM 5.2 was pos/neg;
- TGB, LCA, CD20 were negative;
- Vimentin was positive;
- CD21, CD23, CD35 immunohistochemical markers have not been performed.

EGDS was normal.
Total body CT scan was negative for primary or secondary neoplasm.
MRI of the neck and pharynx, compared to CT scan, reported: “... unmodified bilateral lymph nodes at levels II and V on left side ...”.
It was decided to operate with selective neck dissection (postero-lateral neck dissection on the left side).
During the operation, we partially resected the muscle portion adhering to the neoplasm, since the tumour was adhering in part to the SCM muscle at levels IIA-IIB.

The definitive diagnosis of the surgical specimen was dendritic cell tumour on a 2.7 cm lymph node.
At 18 months’ follow-up, there is no evidence of disease (the patient undergoes ENT and panendoscopy examination once every 3 months and diagnostic examinations – US once every 3 months and total body CT scan once every 12 months).

Discussion
Follicular dendritic cell tumour is a rare intermediate grade malignancy neoplasm with local relapse and metastatic potential, localized – in ENT setting – in the cervical lymph node.
CT scan and MRI are essential investigations in the diagnostic work-up of this disease. Nevertheless, it is only by means of immunohistochemical markers of cytology that we can distinguish between benign and malignant neoplasms and diagnose the nature of the tumour in accordance with histology findings.
There are many pathological conditions that need to be differentiated.
Differential diagnoses with dendritic cell neoplasms particularly interdigital cell tumour/sarcoma are:
- CD21 (receptor of C3d fraction of the complement) is the specific marker of follicular dendritic cells: CD21 is not expressed in Langherans’ cells;
- CD23 and CD35 are specific markers of follicular dendritic cells: like CD21 they are not present in other dendritic cell populations;
- CD1 is negative in follicular dendritic cells but positive in Langherans’ cells;
- S100 protein is a weak and inconstant expression of follicular dendritic cells but is present in tumour interdigital cells (melanoma).
Differential diagnosis with low differentiated carcinomas:
- Anticytokeratin antibodies are epithelial markers which are absent in follicular dendritic cells.
Differential diagnosis with sarcoma:
- Vascular markers (CD31, CD34) and muscle markers (desmin, astin) are negative in follicular dendritic cell tumour.
Differential diagnosis with melanoma:
- S100 protein and HMG-45 are positive markers for melanoma and negative markers for dendritic cells.
Differential diagnosis with thymic tumour (thymoma, thymic carcinoma and Castle):
- Cytokeratin and CD5 are negative in follicular dendritic cells.
The final immunochemical expressive summary of follicular dendritic cells is:
- CD21, CD23, CD35, vimentin: positive markers;
- EMA, S100 protein: pos./neg. markers;
- CD1, cyto keratin, vascular markers, HMB-45, muscle markers are negative.
In the literature, it is reported that the best therapeutic choice for tumours of the neck is surgery. Some reports advise only simple enucleation of neoplasm since pre-operative diagnosis is incorrect; in such cases local relapse or secondary metastasis, after some years, is almost certain.
In agreement with literature, we too have been unable to define, in our case, a right diagnosis of the disease prior to examination of the surgical specimen since the necessary specific immunochemical markers CD21, CD23, CD35 and vimentin (positive in follicular dendritic cell tumour) have not been performed. In our opinion, when the diagnosis is not defined or definable, a surgical neck dissection (radical, modified or selective) would be a better procedure for follicular dendritic cell tumours of the neck.
Radiotherapy or chemoradiotherapy are advisable choices when dealing with particularly voluminous masses, highly aggressive sarcomas, surgically unreachable tumours and when radical surgery cannot be guaranteed.

References


Received October 29, 2003
Accepted March 3, 2004

Address for correspondence: Dr. U. Pignatelli, U.O. di O.R.L., Poliambulanza, Via Bissolati 57, 25124 Brescia, Italy. Fax: +39 030 3515351. E-mail: orl-segretaria.polli@poliambulanza.it