Lymphoma of cheek: a case report


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Abstract. – Lymphoma of cheek is a rare ad uncommon disease, representing 2.5% of malignant lymphoma. The cause is unknown but there are a lot of risk factors such as Helicobacter pylori and Epstein Barr virus. Symptoms are aspecific and may be confused with otolaryngological benign diseases. We present a case of B cell lymphoma of the cheek, which presented with a history of a slowly growing swelling of 3 months duration, resistant to NSAIDs and antibiotic therapy. Biopsy of the mass led to diagnosis of lymphoma. Blood investigations, ultrasonography and CT scan helped to reach this result. This case report shows that an accurate clinical examination, a cytohistological and immune-histochemical diagnosis by fine-needle aspiration biopsy (FNAB) are fundamental to obtain a diagnosis and to decide therapy.

Key Words: Non-Hodgkin’s lymphoma, Lymphoma, Cheek.

Introduction

Non Hodgkin’s lymphomas (NHL) are a group of highly various malignancies and have great tendency to affect organs and tissues that do not ordinarily contain lymphoid cells. 20 to 30% of non Hodgkin’s lymphoma arises from extra-nodal sites. The head and the neck are the second most common region for extra nodal lymphoma after gastrointestinal tract. Nevertheless, primary cheek lymphoma (PCL) is almost rare and really uncommon representing 2.5% of malignant lymphoma.

The cause of PCL likewise the most common NHL is unknown. Currently lymphomas are more likely to develop in immunosuppressive people or in elderly, especially over the 6th decade of life. However, despite the increased risk, NHL is still uncommon in these people.

Certain viruses such as the Epstein Barr virus, which normally causes glandular fever, might contribute to the development of lymphomas and in particular Burkitt lymphoma, an high grade of B cell malignancy. EBV favors the rate of this lymphoma because it has the potential to transform normal human B lymphocytes into growing immortalized cells. It is present in about 50% of Hodgkin’s lymphoma and with varying frequency in non Hodgkin’s lymphomas. One rare type of lymphoma, which usually affects the stomach is known to be caused by a type of bacterial infection known as Helicobacter pylori (MALT = mucosa associated lymphoid tissue). Similar association is known to date with PCL.

Case Report

A 66 years old non smoker, non tobacco addict woman presented with a history of a slowly growing swelling over her right cheek of 3-months duration resistant to NSAIDs and antibiotic therapy. It caused difficulty in opening her mouth in the past 3 months. During this period the patient also accused weight loss, fever, might sweats, weakness and nausea.

The local examination revealed a firm to hard nodular subcutaneous mass on the right side of face reaching up to right lower jaw. The overlying skin was tense and shiny, the mass was well circumscribed and appeared to be free from mucosa and bone. There was no lymph node enlargement.

Her routine blood investigations showed an enhancement of the VES (96 mm 1° hour), C-reactive protein (3.70 mg/dl), alkaline phosphatase (211 v.n. 32-100) and LDH (653 v.n. 250-500). Besides in her CBC it was possible to note an increment of WBC (11,23 × 10³/microL) and neutrophils 85.7% accompanied by an absolute and relative lymphopenia.

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Ultrasonography revealed a hypoechoic well circumscribed mass with the maxima diameter of 4 cm and a laterocervical and submandibular lymphadenopathy. Therefore, patient was subjected to fine needle aspiration biopsy (FNAB) that showed a lymphomatous swelling.

CT scan confirmed the presence of a solid, iso-dense, hypervascolarized mass of 2.5 cm diameter in her right cheek in absence of laterocervical node involvement and metastasis. A percutaneous biopsy was performed in order to obtain an histopathological typing that revealed a small diffuse B cell CD20+ non-Hodgkin’s lymphoma. The immunophenotyping showed B-cell type CD20 and CD79a positive and CD3, CD10, CD15, CD30, CD68 negative. It was also found small lymphocytes with round or cleaved nucleus, minor component of blasts and plasmacytoid cells, compatible with a lymphocyte proliferation.

The patient was considered no operable. Therefore, she received three cycles of chemotherapy (Fludarabine 3.75 mg e.v. days 1-3 + Mytoxantron 15 mg e.v. day 1). This was followed by 18 cycles of local radiotherapy to the diseased area on 6 MeV linear accelerator. She received a total dose of 36 Gy.

Following the treatment there was a partial regression of the tumor but after two months of the regression, patient refused to continue the therapy because of the worsening of performance status caused by pharyngeal infiltration and side effects of radiotherapy (dysphagia, asthenia, paresthesias). Subsequently the patient had an ischiopubic fracture after a long period of bone pains and she died because of acute lung edema.

Discussion

The tumors of oral cavity include several types of malignancies in correspondence of lip, tongue, cheek and pharynx. In the USA, the annual incidence is 19 cases on 100,000 people and the life expectative to five years is about 52%.

Non Hodgkin’s lymphoma is one of the possible cancers in the head and neck region and, between extra nodal non Hodgkin’s lymphomas, this is the second most common site after gastrointestinal tract\(^\text{13}\). In the head and neck, Waldeyer’s ring is the most common site of origin and may be accompanied by cervical node involvement. Nose, paranasal sinus, orbits, salivary gland are other possible organs affected in decreasing order of frequency, with rare spread to the regional lymph nodes\(^\text{14,15}\). Particularly, nasal non Hodgkin’s lymphoma is rare in Western countries but it is very common in East Asian countries and Latin America\(^\text{16}\).

The diffuse large B-cell lymphoma (DLBCL) appears to be the most common type of primary oral and paraoral NHL\(^\text{8,17}\).

There are no characteristics clinical features of lymphoma of the oral region and they depend by the site of the swelling, the lymph node involvement and/or the presence of metastasis. The most common beginning symptoms are local mass, pain or discomfort, dysphagia or sensation of a foreign body in the throat, in the case of tonsillar NHL. It is frequent that a NHL may be confused with a benign disease. Therefore, is useful otorlaryngology examination.

Sometimes, it is also possible found facial hemiplegia and parestesia because of a neural involvement. However, peripheral neuropathy is an unusual complication of lymphoma\(^\text{18,19}\) like so distinct muscular involvement\(^\text{20,21}\). Our patient presented a slowly growing swelling resistant to antibiotics and NSAIDs, weight loss, fever, might sweats, weakness and nausea. Therefore, the symptoms were similar to other types of oral cancers.

The first step, to make a correct diagnosis, is an inspection and palpation of oral cavity and regional lymph nodes.

A part of that instrumental techniques, percutaneous ultrasound (US) and computed tomography (CT) scan are well established procedure. Cytohistological diagnosis is mandatory for diagnosis and treatment with an accurate fine needle aspiration diagnosis of PCL is critical to be realized in time to avoid surgical management and obviates the need for an exploratory laparatomy. FNAB is considered a safe, rapid and easy procedure with high diagnostic accuracy. Percutaneous biopsy should be performed to establish the diagnosis.

Flow cytometry (FC) has significantly enhanced the diagnostic role of FNAB, particularly in the case of hematolymphoid malignancies. FC is extremely sensitive in the detection of antigen expression and identifies small clonal population. FC analysis distinguishes lymphomas from chronic inflammation through the detection of clonality based in surface of Ig light chain expression studies. In lymphomas, IG light chain expression is usually restricted to either kappa or lambda, whereas inflammatory processes reveal a mixed expression of kappa and lambda light chains.
FC also has limited capability in classifying lymphomas into different, well recognized subcategories. This is made possible by studies of surface marker expression and is best applicable for low grade lymphomas. Through this technique it is possible to distinguish malignant lymphomas from non-lymphoid neoplasms, evaluating the presence of positive staining for leukocyte common antigen (LCA). Diffuse large B-cell lymphoma is most commonly positive for CD20 and CD79a and less commonly positive for germinal center markers CD10 and BCL6.

A correct and early diagnosis is important to begin on time therapy. It is easy to change a non-Hodgkin’s lymphoma of the cheek or oral cavity with other benign disease. This causes concern about the performance status and the progression of the cancer. Generally the standard treatment for patients with early stage diffuse large cell lymphoma consists in chemotherapy followed by involved field radiotherapy.

Non Hodgkin’s lymphoma well responds to radiotherapy and the cheek is a region that can be easily treated. However, this therapy is responsible of different side effects like dry mouth, pharynx pain, dysphagia etc. The patient decided to stop the therapy because of the aggravation of performance status also caused by an aggressive therapy.

Patients with advanced stage (bulky stage II, stage III and IV) have to be treated by combined chemotherapy. At this stage the standard treatment should be constituted by 7 cycles of RCHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone).

Our patient received three cycles of chemotherapy (fludarabine 3.75 mg e.v. days 1-3 + mitoxantrone 15 mg e.v. day 1), followed by 18 cycles of local radiotherapy on the interested area by 6 MeV linear accelerator for a total dose of 36 Gy but she decided to stop the therapy for the apparition of side effects depending of radiotherapy.

In conclusion, non Hodgkin’s lymphoma should be considered in the differential diagnosis of various benign and malignant lesions because the treatment and prognosis are very different in the several cases. However, according to the international prognostic index (IPI), established for patients aged less than 60 years, the outcome of patients with extranodal DLBCL is similar to that of patients with nodal DLBCL. Between malignant diseases we have to consider the presence of different histopathological cancers such as liposarcoma, carcinoma, Sarcoma of Kaposi and Hodgkin’s lymphoma.

Finally, an accurate clinical examination, a cytohistological and immune-histochemical diagnosis by FNAB and flow cytometry became fundamental steps to decide a proper therapeutic protocol.

References
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