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Primary Cutaneous B-Cell Centrofollicular Lymphoma: Role of Surgery

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Abstract

Primary Cutaneous Follicle Center Lymphoma (PCFCL) is a big entity of a large group of B cell lymphoma. In this paper, we report a case of (PCFCL) with particular clinical presentation (huge mandibular mass) successfully treated by a surgical approach followed by R-CHOP chemotherapy. The aiming of our article is to highlight the role of surgery in the management of this group of tumors. Our case concerns a 47 years old female patient, admitted for a large red tumor in her right lower cheek. We performed a surgical resection with 1 cm marges, followed by a reconstruction with a major pectoralis flap. The histopathological and immunohistochemical came back in favor of centrofollicular B cell cutaneous lymphoma. The patient then benefited from 2 cures of R-CHOP chemotherapy followed by 3 cures of R.

Subject Areas

Surgery & Surgical Specialties

Keywords

Primary Cutaneous Follicle Center Lymphoma, Role of Surgery, Major Pectoralis Flap, Case Report

1. Introduction

Primary Cutaneous Follicle Center Lymphoma (PCFCL) is a low lying B cell lymphoma grade which generally affects adults in their fifties [1], even if cases can be seen in young or older subjects. The lesions are papules or fixed erythematous nodules, willingly multiple, although often grouped together in a restricted cutaneaous territory, preferentially affecting the cephalic extremity and the trunk. It is an indolent lesion, with a 5 years survival rate of 95% [2].

We report a case of PCFCL with particular clinical presentation (huge mandibular mass) successfully treated by a surgical approach followed by R-CHOP chemotherapy.

The purpose of this work is to demonstrate that surgery could be an effective way to treat PCFCL without being too aggressive as in chemotherapy/radiation treatments.

2. Case Report

We report a case of 47 years old female patient with a known history of chronic polyarthretis rhumatoid under methotrexat, admitted in oral and maxillofacial surgery department for a large red tumor in her right lower cheek gradually increasing in size for 2 years, clinical examination showed a 10 cm red purpilish mass localized in the right lower cheek area, with smooth irregular surface, with a good peripheral vasculirazation, mobilased relatively to underlying planes, in particular the mandibular one.

In front of this clinical picture (**Figure 1**), we suspected a vascular tumor, an angio MRI was indicated showing a massive round tumoral process with a polycicls countours, fairly limited on hyposignal T1 and intermediate signal T2, infiltrating the adjacent soft part without cortical bone invasion (**Figure 2**, **Figure 3**).

Absence of cercival lymph node.

As far as the angio MRI was not concluding, we completed with a carotid selective arteriography which showed no abnormalities.

Based on radiological results, the patient benefited from surgical resection by a maxillofacial qualified head attending professor, under general anesthesia with orotracheal intubation, we performed a large tumoral resection with 1 cm marges, the soft and bone parts were found intacts, without peroperatory bleeding, a reconstruction with a major pectoralis flap was done (**Figure 4**).

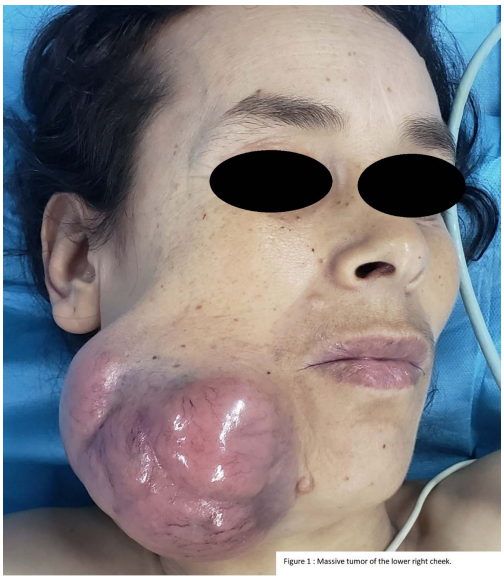

Figure 1. Massive tumor of the lower right cheek.

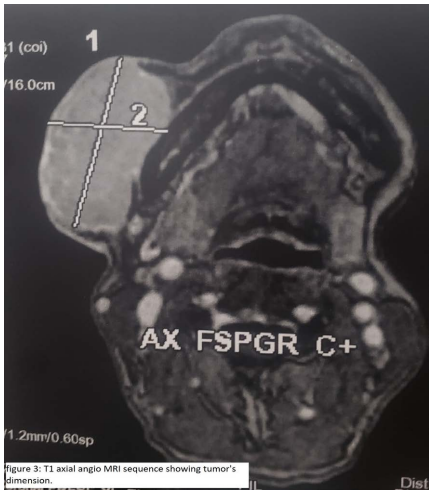

Figure 2. T1 axial angio MRI sequence showing tumor's dimension.

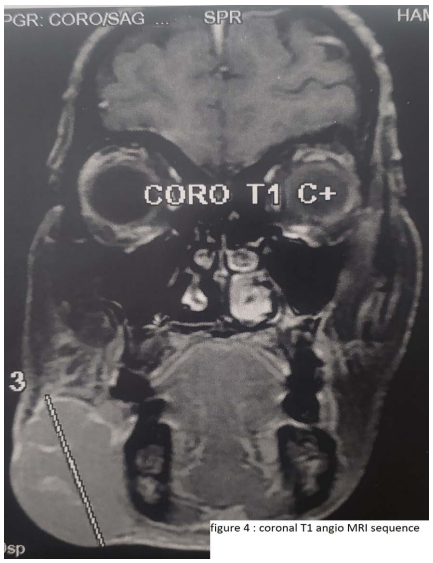

Figure 3. Coronal T1 angio MRI sequence.

Figure 4. Post-operative presentation.

The histopathological and immunohistochimical results resulted with centrofollicular B cell cutaneous lymphoma.

the samples taken of the tumor show a cutaneous tissue made of a regular epidermis separated from the superficial dermis by a band under the epidermis, the underlying dermis is the seat of a malignant tumoral proliferation reaching the hypodermis of nodular architecture made up of cells of large, atypical sizes, with a cleaved and elongated nucleus surrounded by a cytoplasm of a clear appearance, several figures of mitosis are noted.

Presence of tumor necrosis.

The stroma is fibro-inflammatory.

An immohistochemical complement was carried out showing a positive labeling of the tumor cells by CD20, BCL2, BCL6 and CD10.

Labeling of reactinnel T lymphocytes by CD3.

Absence of marking of tumor cells by CK20.

The Ki 67 proliferation index is estimated at 70%.

A body scan was recurred for extension evaluation that came back negative with no radiological significated abnormalities; based on this finding, the diagnosis of primary cutaneous centro follicular B cell lymphoma (PCBCL) was established.

The patient was then addressed to oncolongy unit for 2 cures of R-CHOP chemotherapy (Rituximab (375 mg/m^2), cyclophosphamide (750 mg/m^2), doxorubicin (50 mg/m^2), vincristine (1.4 mg/m^2), and prednisone (100 mg/j) followed by 3 cures of R (Rituximab (375 mg/m^2)).

The patient is successfully on remission after a follow up of 12 months; a regular control every 3 months is being set up; no sign of recurrence is noted.

3. Discussion

PCFCL is more common among men than women with a median age of 50 [1], It's part of a large group of primary cutaneouas B cell lymphoma (PCBCL), having a slow growth process, it has an excellent prognosis (PCFCL) is often localized in neck, head area with a percentage of 61%. 20% have a unilesional presentation while 80% have multiples lesions [1] [3].

Extracutaneous spread is rare less than 10% [1].

In general, (PCFCL) has slow-growing that takes years with a quite smooth surface, 2 cm to 5 cm, they can be presented with many forms: nodules, plaques, or tumors. These usually have telangiectasias [1] [3]. In our case the patient presented a 10 cm tumor that occurs in an lower right cheek with a quiet smooth thelengisctasic surface, the growth process took approximatively 2 years, our results add up with the literature description, however the unusual part was the huge size of tumor that leads to believe more of a vascular malformation.

The histology of PCFCL is variable. The infiltrate is made up in variable proportions of small regular lymphocytes, part of which is composed of reactive T cells, and of cells reminiscent of the elements constituting the normal germinal

center of activated lymphoid follicles, which represent the "normal counterpart" of cancerous cells. Cancer cells are thus B lymphocytes, of centrocytic or centroblastic morphology, constantly expressing the BCL6 marker and, more inconsistently, CD10. These cells form either diffuse layers or, more rarely, nodules, reminiscent of the appearance observed in lymph node follicular lymphomas, which belong to the same family [4] [5].

Most PCFCL have immunophenotypical profil that includes CD 19, CD 20, CD 79a+, and PAX 5 positive markers, plus at least one follicule center marker, that is often BCL6, less likely CD 10, Ki67 is a mitotic index, that is associated with cellular proliferation, it helps determine whether it is a follicular reactive hyperplasia and follicular malignancies [6] [7].

In most cases PCFCL cells don't/rarely express BCL2 marker, however in our case we occured a positive marquage of neoplasic cells with BCL2 marker [8] [9].

The treatment of PCFCL is important because neglected forms can spread, the different therapeutic modalities consist of surgical resection, radiotherapy and chemotherapy [10].

Surgical excision may be indicated for solitary, small and well defined lesions, however, the resection margins remain unknown, the recurrence rate is estimated at 33% of cases [11].

Unfortunately, few studies have been dedicated to highlight surgery role in (PCFCL), in Sam Parbhakar *et al.* study, it has been suggested that surgical excision should be considered as a first line treatment in marginal zone and follicle centre subtypes, given that it prevents local complications that could be associated with radiotherapy.

In literature reviews radiotherapy is a therapeutical option for (PCFCL) [12].

In multifocal, extensive skin lesions, aggressive forms and extra-cutaneous manifestations chemothearpy that associate cyclophosphamide, hydroxydoxorubicin, vincristine, prednisone, and rituximab (R-CHOP) is a option that should be considered [7] [13].

4. Conclusion

Surgical excision remains the main option for primary cutaneous centrofollicular B cell lymphoma combined with chemothrapy/radiotherapy for optimal results in some cases.

Conflicts of Interest

The authors declare no conflicts of interest.

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