

Tumefacção parotídea: Primeira manifestação de linfoma de Hodgkin

Parotid mass: The primary manifestation of Hodgkin's lymphoma

Antonio Caravaca-García • Rafael Franco • Rosario Guerrero-Cauqui • Rafael Ruiz-Fito

ABSTRACT

The finding of lymphomas in the salivary glands is uncommon. Moreover, the diagnosis of Hodgkin's lymphoma from the finding of an initial swelling that is increasingly growing in the parotid region is rare. The clinical examination, the FNA or the radiology investigations including CT scan, will hardly help in understanding the nature of the disease. Because of this, lymphoma must be taken into account in the differential diagnosis of parotid masses.

Keywords: Hodgkin's lymphoma, parotid mass, salivary glands.

INTRODUCTION

Lymphoma is a disease of the lymphatic system that can be classified into two categories: Hodgkin's Lymphoma (HL) and non-Hodgkin's Lymphoma (NHL). These can be sub-classified in different subtypes. Although most lymphomas are located in the lymphatic ganglia, others may appear in different organs, manifesting themselves as an extra nodal disease^{2,4}: approximately between the 25-30% of the cases of NHL and 1% of the HL¹³. The gastrointestinal tract and the head and neck are usually the most frequent sites of presentation⁴.

Parotid gland lymphomas are uncommon^{1,4,7,12}, representing 0.6-5% of all these type of tumours^{1,7}. Some authors have described an increased frequency of the parotid lymphomas⁸. Among them, the immense majority are NHL, being anecdotal the presentation as HL form. The case reported herein is a long standing parotid lymphoma in a 26 years-old man. The differential diagnosis, the treatment and the management of this type of tumour are discussed.

CASE REPORT

A 26 years-old man was sent to the ear-nose-throat (ENT) department at the Hospital, complaining of a small swelling in left parotid area which remained asymptomatic during the last few months. However, in the last weeks he noticed an increased size of the mass. The physical examination showed a uniform mass of about 2 cm of diameter in the middle of the parotid gland. There were not cervical lymphadenopathies and the facial nerve remained intact. The rest of the examination of the head and neck was normal.

(Figure 1)

Among the investigations the FNA, that was previously performed, led to the first diagnostic approach of Warthin's tumour. The magnetic resonance image (MRI) displayed an intraparenchymal mass of about 2 cm of diameter in the superficial lobe of the parotid gland (Figure 2).

Surgical intervention was suggested to the patient, who accepted and signed the consent form. Surgery was

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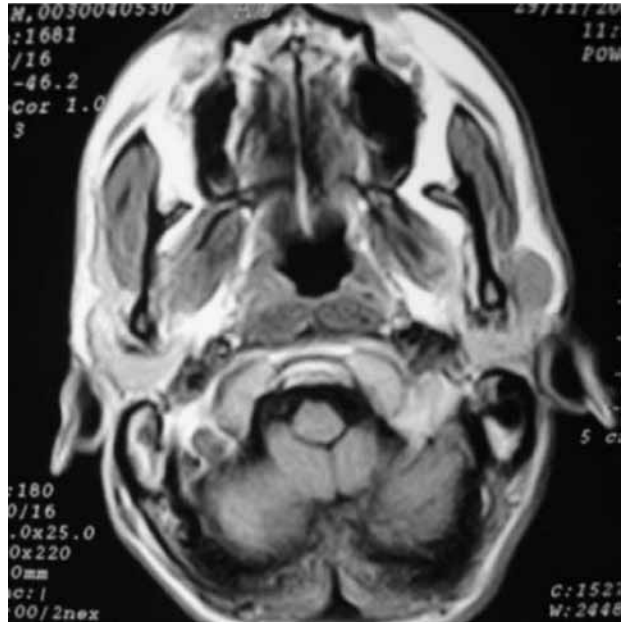
FIGURA 1

Lateral view of the patient face



FIGURA 2

Axial of MRI: the lesion into the parotid gland



carried out by means of a superficial parotidectomy with preservation of the facial nerve (Figure 3). No more lesions were detected.

The histological sample was sent to the National Centre of Oncological Research (Madrid), which confirmed the diagnosis, corresponding to the subtype of a "Nodular lymphocyte-predominant" being negative for CD 15, CD 30 and P53 CD and presenting a low proliferative index. (Figures 4 and 5)

The patient was referred to the Haematology department of our hospital, where the guidelines

were carried out which included CT SCAN of abdomen and chest and a PET, confirming the absence of other regional involvements. The final diagnosis was stage I EA without unfavourable prognosis.

Radiotherapy was administered to the affected zone, by means of 40 Gy, distributed in a 2 Gy dose by session and 5 sessions per week during four weeks. The only side-effect was first degree radiation-induced dermatitis.

FIGURA 3

The parotid mass during the surgery

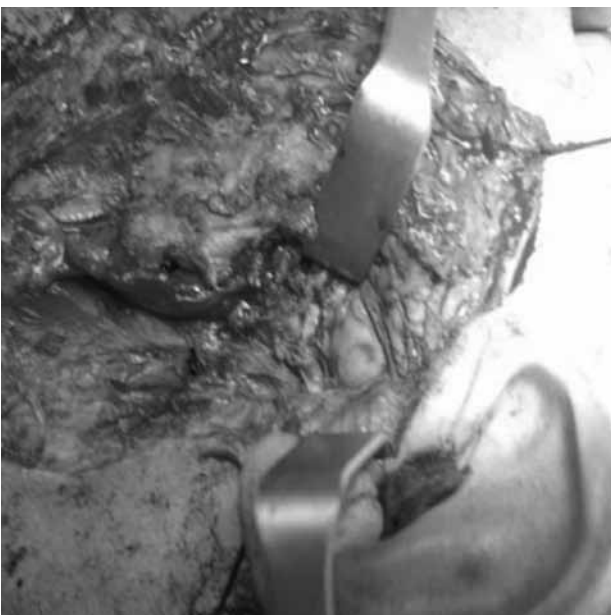


FIGURA 4

Hematoxylin eosin staining shows glandular tissue (on the right) of normal features, which surrounds the lymphoid growth which architecture is locally modified by proliferation of a **nodular pattern lymphoid neoplasia**.

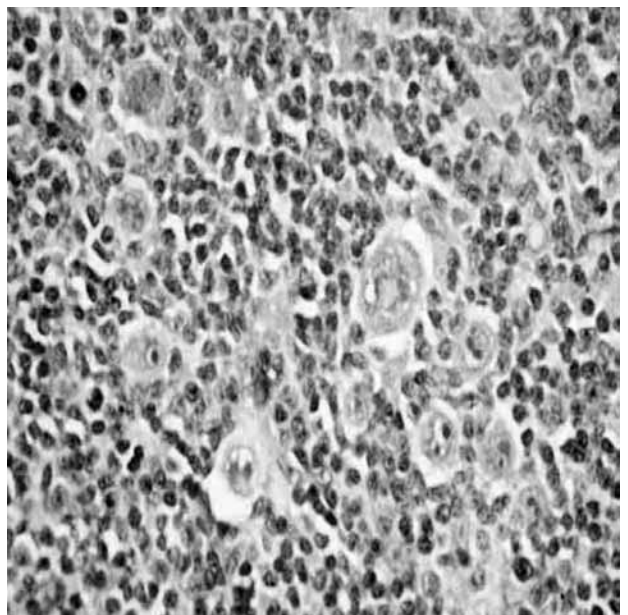
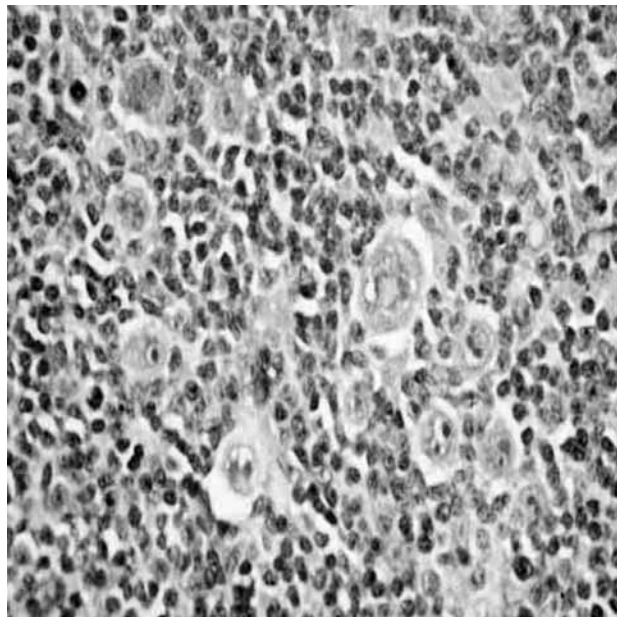


FIGURA 5

Lymphoid population is polymorph and **Reed-Stenberg** cells are found in the lymphoid follicles and interfollicular areas; these cells are loose, big, polinuclear and have a wide cytoplasm



The postradiotherapy MRI and PET which were performed after the treatment did not show any lesion, and the treatment was over.

DISCUSSION

The HL is around fivefold less frequent than the NHL (1). It has a bimodal distribution⁴, with an average age appearance of 32 years. Risk factors for developing lymphoma are: immunodeficiency (acquired or congenital), previous radiation, immunosuppression, Sjögren's Syndrome, rheumatoid arthritis, celiac disease and thyroiditis of Hashimoto.^{1,4}

From a pathology point of view, the HL is classified in two types (Table 1)³: Nodular lymphocyte-predominant Hodgkin's lymphoma - NLPHL - (5%) and the Classical form - CHL - (95%) which is subdivided in Nodular sclerosis classical Hodgkin's lymphoma (that supposes a 70%), Mixed cellularity classical Hodgkin's lymphoma (around a 25%), Lymphocyte-rich classical Hodgkin's lymphoma (a 3%) and Lymphocyte-depleted classical Hodgkin's lymphoma (a 2%). The case corresponds to the first described type, a rare organization in contrast

TABLE 1

Who classification of Hodgkin's lymphoma⁽³⁾

- Nodular lymphocyte-predominant Hodgkin's lymphoma
- Classical Hodgkin's lymphoma.
 - o Nodular sclerosis classical Hodgkin's lymphoma
 - o Mixed cellularity classical Hodgkin's lymphoma
 - o Lymphocyte-rich classical Hodgkin's lymphoma
 - o Lymphocyte-depleted classical Hodgkin's lymphoma

to the more frequent classic forms.

The morphologic characteristics of the NLPHL consist on the presence of lymphocytic cells and atypical histiocytic cells ("pop corn" cells) which are surrounded by a substructure of follicles in progressive transformation, being characteristic the little presence or absence of Reed-Sternberg cells, as it happens in CHL type^{3,4}.

Lymphomas that appear outside the lymphatic ganglia denominates themselves extranodals, as they are located in the parotid gland^{1,4}, being able as well to originate themselves in the intraparenchymatous ganglia or the own glandular tissue. They can also be classified as primary or secondary, following the different criteria used in medical literature. (Table 2)⁴

TABLE 2

Criteria used to define primary parotid gland lymphoma⁽⁴⁾

- Any lymphoma originating in either glandular lymph nodes or glandular parenchyma, regardless of its association with autoimmune disease or its subsequent stage; must be initial clinical manifestation of lymphoma..
- Any lymphoma originating in glandular parenchyma as initial clinical manifestation of lymphoma.
- Any lymphoma not associated with autoimmune disease, benign lymphoepithelial lesion, or myoepithelial sialadenitis.
- Any lymphoma originating in glandular parenchyma not associated with autoimmune disease.
- Any lymphoma originating either in glandular parenchyma or in glandular lymph node with invasion of parenchyma, in absence of detectable disease outside of parotid gland.
- Any lymphoma originating in glandular parenchyma in absence of extraparotid involvement at time of diagnosis.

The parotid lymphoma usually appears like a long standing asymptomatic mass^{4,7}, indistinguishable from any other tumour of epithelial origin⁷. Although the FNA is a standard test in managing the diagnosis, its accuracy varies from 66 to 93%. Because of this, there appears a high frequency of misdiagnosis and confusion with Warthin's tumour. The definitive diagnosis will be made almost always after an untimely histological study of the piece of parotidectomy and its phenotypic determination^{1,4,7,8}.

The primary HL in parotid gland is extremely rare (few cases described in literature⁴⁻⁵⁻¹⁰ the commonest one being the predominantly lymphocytic classic form of the HL.

Parotid lymphomas have a favourable prognosis, much more than the rest of lymphomas in general, and the HL has a better one than the intraparotid NHL: with a 5 years survival rate of 90% as opposed to an 80% of the latter (4). The age and the stage at the time of diagnosis have more importance in the prognosis than the histological subtype.

Although it is possible the diagnosis by means of FNA, the surface parotidectomy is recommendable^{1,9,10}, being used more and more the monitoring of the facial nerve⁶. The radiology investigations are CT SCAN or MRI and they are not definitive in the diagnosis, although they are included in the preoperative investigations^{1,4,6,7,9}. Pathognomonic data do not exist: The CT SCAN usually shows a unilateral and poorly defined soft mass, with homogenous tissue, with little differentiation of the margins of the tumour¹¹. The staging will include a later CT SCAN of pelvis, abdomen and chest^{3,4,7}, being the haematology and radiotherapy specialists the ones in charge of the treatment with chemotherapy and/or radiotherapy^{1,3,4} according to the staging.

CONCLUSION

The parotid primary HL has a rare organization, that usually is not suspected in the initial assessment of a parotid mass. The most frequent subtype is the predominantly lymphocytic type of the CHL, representing a peculiarity the presented case. Its prognosis is more favourable, with a 5 years survival rate of around 90%. Its treatment includes chemotherapy, radiotherapy or both.

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