

Orbital Lymphoma: About a Case Revealing Diagnosis Challenges

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ABSTRACT

Orbital lymphoma is rare by its localization. It's usually a B cell lymphoma. The diagnosis relies on the histological study after a biopsy. The biopsy could be a challenge depending on the area of the tumor and the anatomical structures near the lesion. In the article we are presenting a case of patient with a tumor located in the orbit. Usually, the biopsy is made through the skin. The endonasal way for a biopsy of an orbital tumor can be more useful and safer than a classic way through the skin. Here are an article describing a case of orbital lymphoma, management and the characteristics of the pathology.

Keywords: Lymphoma, Orbital, exophthalmia, rhinosinusitis.

Case Report

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INTRODUCTION

Orbital lymphoma is a rare entity, and it concern especially the B cell lymphoma type. The diagnosis relies on the histopathological examination as any tumor. Clinical signs are common between the orbital tumors, and not specific. There is various differential diagnosis regarding the site of the lesion. A whole body exploration is necessary to find out if there are other localizations before starting the treatment.

Orbital tumors need to be well evaluated before by non-invasive tests. Biopsy or fine needle puncture could be harmful through palpebral skin.

In this paper we expose a case of patient with orbital lymphoma, the diagnosis was confirmed after a biopsy through endonasal technic respecting the palpebral skin.

CASE:

It's about a 39 years old woman, with exophthalmia appeared 1 month before the consultation in the left eye (Figure 1). There was no diplopia reported and no rhinosinusitis dysfunction. The endonasal examination shows an edema in the middle meatus in the same side of the exophthalmia.



Figure 1: Exophthalmia of the left eye

The CT-scan imaging shows a tumoral lesion invading the orbit with an extension through the nasal cavity invading the middle meatus (Figure 2).

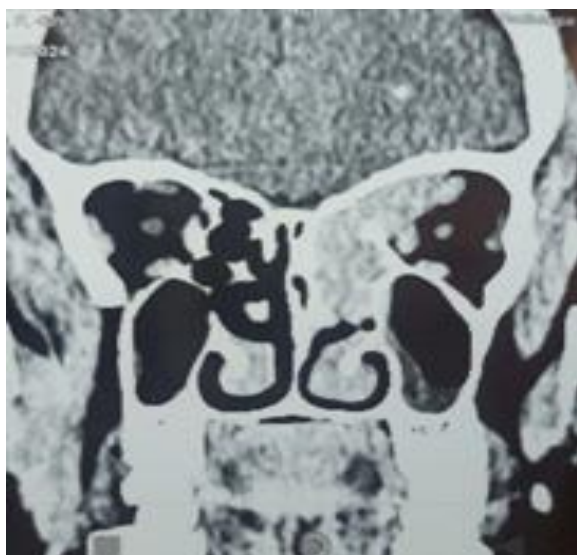


Figure 1: CT- Scan shows the extension to the nasal cavity

A biopsy was done by using endoscope introduced in the left nasal cavity. After using a Xylocain with Naphazolin for infiltration in a goal to reduce bleeding, we took multiple fragments from the mass.

The histological examination and the immune histochemical tests objective a B cell lymphoma: Ki 67 positive and CD20 positive (Figure 3 and 4).

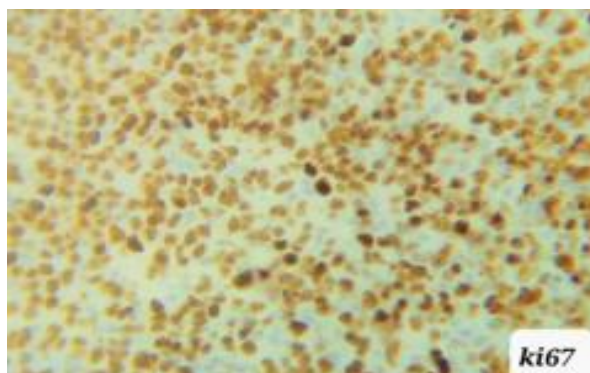


Figure 3: Ki 67 positive

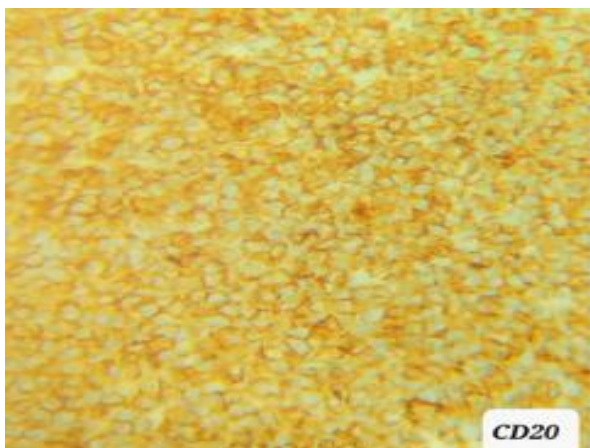


Figure 4: CD 20 positive

A treatment by radiotherapy was decided after multidisciplinary reunion.

DISCUSSION

The orbital lymphoma is for most cases a B cell lymphoma. It grows spontaneously in the anatomical spaces and produces a mass effect on the structures next to the tumor. The orbit is shaped as a quadrilateral bony structure that contains the eyeball and structures of the ocular adnexa [1]. Its medial wall is a bony structure, the lamella papyracea which is thin and can be easily destructed.

The development of a mass in such narrow structure could lead to engage the functional prognosis of the eye. That depends also on the rapidity of the growth and its type, if it's malignant or benign. Mostly malignant tumors are responsible of bone lysis. Usually, the lamella papyracea is frequently concerned, due to its reduced thickness. So, an adnexal orbital tumor which rises in the medial side of the orbit could extend in the homolateral nasal cavity. Frequently it will be an exophthalmia due to the mass effect. Some cerebral tumors can have a similar aspect such as glioma or meningioma [2].

The symptomatology is not specific and it could be pain, diplopia and swelling, sometimes it's just an exophthalmia reported by the patient himself. The diagnosis is done with a CT-scan and a biopsy. Most of the time the biopsy is done depending of the localization of the tumor, it can be a blepharoplasty incision or a lateral orbitotomy [3]. But the biopsy site depends especially on the localization of the tumor. It's not simple to have an endonasal access for it, if the site is in the supero-lateral quadrant as it's described most frequently [2]. It can involve each of the superior rectus muscle, lateral rectus muscle, lacrimal gland and eyelid [2]. Or if the tumor is limited and not big enough to an endonasal access. The endonasal way is clearly the best way to get that fragment for histological study. Adding the fact that the inflammation generated by the incision can participate in the diffusion and the spreading of the tumor. The orbit is almost full with the tumor and there is no risk to expose orbital structure to compression with an hematoma after a blepharoplasty for example.

The only way to get a certain diagnosis if the histological examination. Extranodal marginal zone B-cell lymphoma (EMZL) was the most frequent subtype [4]. The surgical biopsy could be inesthetic and the scar left behind can participate in the diffusion of the lesion and make it more difficult for treatment and expose the patient to serious complications after radiotherapy [5].

Radiotherapy is recognized till now as the most efficient treatment for orbital lymphoma [6].

CONCLUSION

Orbital lymphoma is mostly a B cell lymphoma. The treatment relies on radiotherapy. The biopsy could be the only surgery needed. The necessary time for healing can delay the time to start the radiotherapy and even the scar left of the palpebral skin can could complicate the radiotherapy. That's why we encourage endonasal biopsy if it's possible.

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