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Orbital Lymphoma on a Case in a Secondary Center in Sikasso

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Abstract **Case Report**

Malignant lymphomas constitute a very heterogeneous group of malignant proliferation of lymphoid tissue whose starting point is extramedullary. The confirmatory diagnosis is based on the anatomopathologic examination of the surgical specimen. Immunohistochemical typing is used to determine Hodgkin's Lymphoma (HL) and other non-Hodgkin's lymphomas (NHL). The treatment of these lymphomas involves a multidisciplinary approach and includes surgical excision, cryotherapy, chemotherapy which can be associated with radiotherapy. Very few studies carried out on orbital lymphomas in Sikasso. The purpose of our study was to write the diagnostic procedures in a secondary center and the management as well as the prognosis. It was a 23-year-old young woman with no particular history, no notion of taking stimulants, who consulted for a necrotizing and painful swelling of the right eye evolving for more than 1 year in a progressive manner despite corticosteroid therapy. Imaging and pathological examination of the surgical specimen led to the diagnosis of large cell lymphoma. Surgical excision was the therapeutic means performed. The postoperative course was good with healing of the wound after 2 months. After 3 months of follow-up, the patient would have died after a period of loss of sight. The clinical polymorphism of lymphomas can mask the diagnosis and delay the therapeutic management which determines the prognosis.

Keywords: Orbital swelling, orbito-cerebral scanner, large cell lymphoma.

INTRODUCTION

Orbital localization of lymphoma is rare [1]. Malignant lymphomas constitute a very multifaceted group of malignant proliferation of lymphoid tissue whose starting point is extramedullary. These lymphomas have very diverse and varied biological characteristics (immunophenotyping) with a possible role of exogenous factors (infectious agent) or endogenous factors (an autoimmune disease) [2]. In general, lymphomas include Hodgkin's lymphoma (LH: 15%) and other non-Hodgkin's lymphomas (NHL: 85%) developed at the expense of B lymphocytes (which produce antibodies) or T lymphocytes (responsible for immunity cellular) [2].

They represent 5 to 10% of tumors of the appendages of the eye and 8% of orbital tumors [3, 4]. These lymphomas frequently occur in elderly subjects around 60 years old and are almost always of high malignancy, especially when they concern young people.

The treatment of these lymphomas involves a multidisciplinary approach and includes surgical excision, cryotherapy, chemotherapy which can be associated with radiotherapy.

Anatomopathological examination makes it possible to make the diagnosis. We report the case of a 23-year-old female subject in the Sikasso region. The aim was to write the diagnostic procedures in a secondary center and the management as well as the prognosis.

OBSERVATION

It was about a young woman of 23 years who consulted for a painful swelling of the right eye evolving for more than 1 year in a progressive way despite corticosteroid therapy. On examination, there was a significant, very inflammatory swelling with pain that interfered with sleep. On palpation, the swelling was hard and immobile with a completely necrotic eyeball and visual acuity was limited to an absence of light perception for Right Eye (OD) and 10/10 for Eye (OG).

In front of his signs, a Maxillofacial opinion and an orbito-cerebral scanner were carried out. Thus the maxillofacial examination objectified and confirmed the extension of the tumor at the level of the mouth (Fig 5). Orbito cerebral computed tomography (CT) examination showed orbital swelling and retro orbital with maxillary and right sinus extension (Fig. 4). Faced with the persistence of insomnia pain, a preoperative assessment was carried out. After a multidisciplinary consultation, a biopsy plus surgical excision was decided and performed in collaboration with the maxillofacial surgeon. The approach was orbito-maxillary (Fig 3) using the Weber-Ferguson technique, which allowed us to perform enucleation first (Fig 8) and then excision of a large bleeding tumor (Fig 9). Histological examination of the specimens revealed a large cell lymphoma. We then did not perform immunohistochemical typing and other extension assessments for economic reasons and a limited technical platform at our center.

Our patient was followed until complete healing of the surgical wound in the ophthalmology and maxillofacial department (Fig 6 and 7) for 2 months. For lack of financial means, the extension assessments given to the patient could not be carried out in a tertiary center in Bamako. A monthly monitoring of 3 months was instituted after which, she was lost sight of and she would have died 3 months later. The failure to carry out remote extension assessments was an obstacle to making the link between his tumor and the causes of his death.

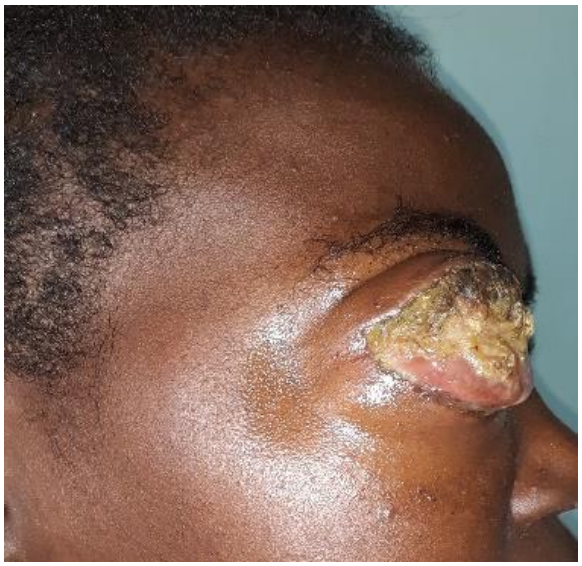

Fig 1: Necrotic globe with a protrusion

Fig 2: Necrotic globe with a protrusion

Fig 3: The maxillary orbit approach (Weber-Ferguson technique)

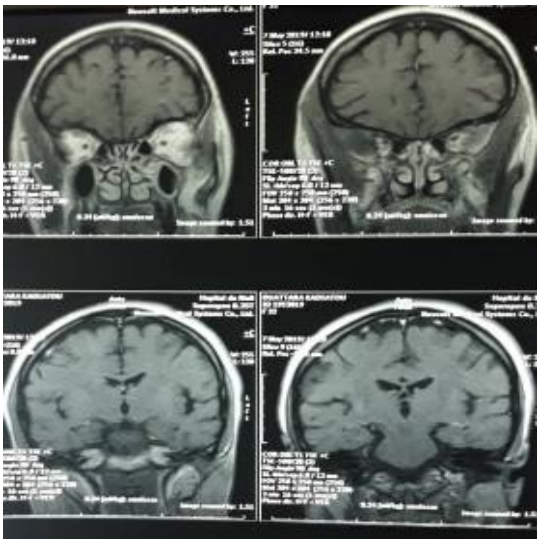

Fig 4: Orbito-cerebral CT scan

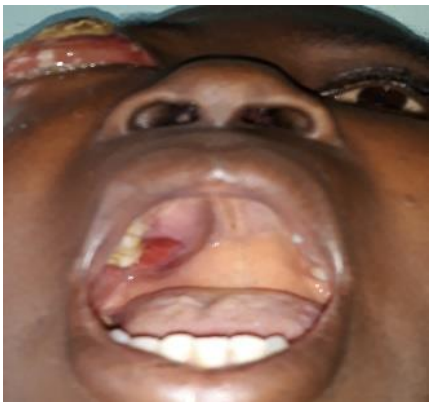

Fig 5: Buccal extension of the tumour

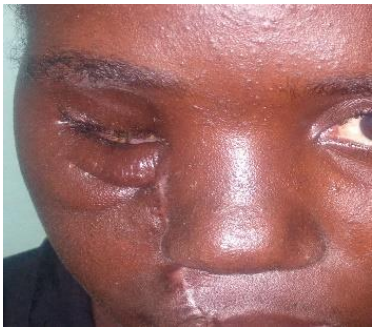

Fig 6: Healing of the surgical wound 2 months after excision of the lymphoma

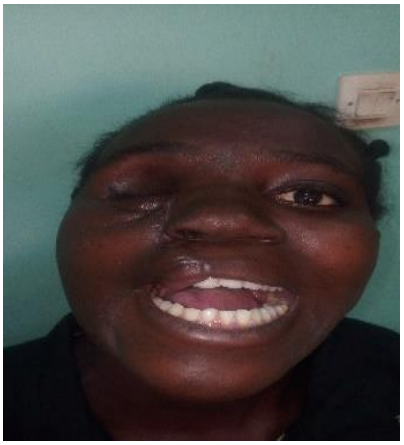

Fig 7: Healing of the surgical wound 2 months after excision of the lymphoma

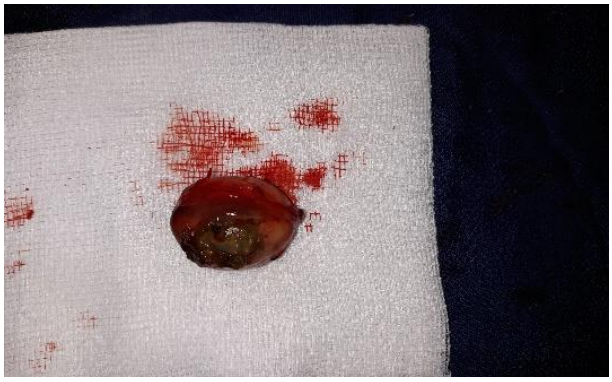

Fig 8: Excision of the eyeball

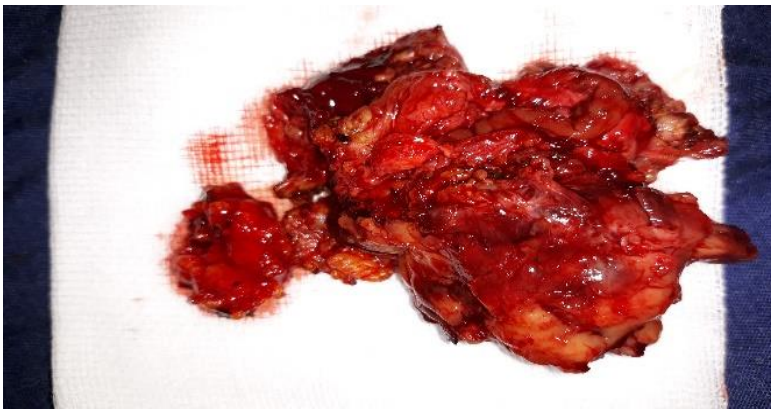

Fig 9: Excision of the large bleeding tumor

DISCUSSIONS

Lymphomas of the orbit, although rare, represent less than 5% of non-traumatic orbital diseases [1]. Orbital lymphomas comprise 8% of orbital tumors [4]. The clinical symptomatology of lymphomas is very diverse and varied. In our study, the age of our patient was 23 years old, close to the average age found by SYLLA F at the CHU IOTA in Mali and by SANOUSSI S in Niger who respectively found an average of 32.19 in a series of 63 cases. tumors and 29.49 in a series of 27 cases [5, 6]. On the other hand, it was much lower than that of Vasseur and Bouchama who respectively found an average age of 40 and 63 years in France and Morocco [7, 8]. Our study concerned the subject of female sex, similar to the study of SANOUSSI S in Niamey and L Benabid in Amiens in France who respectively found 27 cases and 22 cases with a female predominance each [3, 6]. BOUCHAMA in Morocco reported this female predominance with a sex ratio of 1.5 or 6 women out of 10 cases [8], on the other hand AY SEGBENA in Togo found a predominantly male sex ratio in a series of 28 cases, i.e. 17 patients (60.7%) male and 11 (39.3%) female [9], R JOUBERT in France had found a parity between the two sexes in his series of 6 cases [10]. In our observation, the presenting sign was painful necrotizing exophthalmos (Fig 1 and 2), on the other hand AY SEGBENA in Lomé found exophthalmos at 42.9%. in his series of 28 children [9]. Orbital swelling and retro-

orbital with maxillary and right sinus extension was the sign of regional extension, observed by the maxillofacial and confirmed by CT scan. We do not have a PET-scanner or an MRI at our centre.

For management, we performed a biopsy-exeresis with the orbito-maxillary approach using the Weber-Ferguson technique which consisted of an incision by lateral rhinotomy, a labial and lower palpebral section (Fig 3). Roussel in France and YACOUBI B in Algeria respectively used excisional biopsy of the lacrimal gland with a transseptal anterior orbital approach [1] and a monobloc fronto-orbital approach. They thus proceeded with a large excision of very invasive tumors, very hemorrhagic, soft, friable, without clear limits with the orbital contents [11]. The choice of our technique was related to the clinical aspect (necrosis and pain) as well as to the locoregional extension of the tumour. On the other hand, in the series of L Benabid, the symptomatic forms or of high malignancy were treated by chemotherapy associated with localized radiotherapy in first intention with new therapies which seem to be effective in lymphomas of low malignancy, such as the use of anti-CD20 antibody or immunotherapy [3].

The confirmatory diagnosis was made by the pathological examination of the cells of the operating specimen, which corroborates well with the study of the clinical cases of Roussel in France and H. El Maaroufi in Morocco [1, 12]. In addition, in the series of AY SEGBENA, 22 patients (78.6%) out of 28 cases, benefited from the anatomopathological examination and the cytological analysis [9]. The presence of large cell lymphoma on the anatomopathological examination in our patient was also found by SZWARCBERG J in Strasbourg [13] in malignant phenotype B lymphoma, which was partly large cell. We were limited in carrying out remote extension assessments and typing. Examination of the results of the extension assessments and that of the clinical and radiological assessments and the anatomopathology should make it possible to classify the lymphoma as benign or malignant (low grade or high grade) and this to guide the post-surgical treatment protocol.

Our patient was lost to sight after 5 months of follow-up, L. Benabid and recorded 5 lost to sight (3 deaths and 2 without information) out of 22 cases [3]. All 28 cases in the AY SEGBENA study were lost to follow-up after discharge from hospital. Only one patient among the 18 was followed for 18 months before being lost to follow-up [9]. Our patient died 6 months after the healing of her surgical wound, this delay is less than that found in the study of R JOUBERT in France, with a survival period of 12, 18 and 24 months of the patients followed [10]. This difference could be explained by the quality of the technical platforms, the early diagnosis of the disease and the quality of care.

CONCLUSION

The clinical presentation of lymphomas is very variable and depends above all on the site of development of the tumour. Exophthalmos is the most frequent warning sign. The clinical polymorphism of lymphomas can mask the diagnosis and delay the therapeutic management which determines the prognosis. The key to diagnosis remains the pathology study.

When the financial means and diagnostics are lacking, surgical excision can be considered as first intention followed by a treatment adapted to the clinical condition of the patient when the globe is irrecoverable with intense pain.

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