

Orbital Metastasis of a Pediatric Nasopharyngeal Carcinoma: A Rare Case Report

M. Guezar^{*1}, S. Khalfi¹, MA. Erraïsse¹, M. Iken², M. Boubou², FZ. Farhane¹, Z. Alami¹, T. Bouhafa¹.

⁽¹⁾ Radiation Oncology Department, Oncology Hospital, Hassan II university Hospital, Fez, Morocco

⁽²⁾ Radiology Department, Hassan II University Hospital, Fez, Morocco

*Corresponding author : M. Guezar¹

Abstract:- Although nasopharyngeal carcinoma (NPC) has a high tendency for early distant spread, orbital and ocular adnexa metastases are rare. This paper reports a 10-year-old female child previously diagnosed with a locally advanced NPC, she was treated with neoadjuvant chemotherapy based on bleomycin, epirubicin, and cisplatin (BEC) protocol with a low radiological response. Afterward, the management was supplemented by concomitant chemoradiotherapy. The patient subsequently presented bilateral painful palpable masses in the medial canthal area with mild proptosis, redness, and decreased visual acuity. The cervical facial scan showed bilateral tissue formations of the internal canthus, related to secondary locations. The cerebral scan showed a tissue formation of the frontal bone, enhanced after contrast without periosteal reaction, related to a secondary location.

Subsequently, the patient received two cycles of palliative chemotherapy. The evolution was marked by the progression of orbital metastases. The patient died two months after the diagnosis of orbital metastases. In this case report, we illustrate a new case of a pediatric nasopharyngeal carcinoma with orbital metastasis and report the clinicopathological features and therapeutics interventions, as well as the outcomes of this aggressive disease.

Keywords:- Orbital Metastases, Nasopharyngeal Carcinoma, Radiotherapy, Chemotherapy, Imaging.

I. INTRODUCTION

Orbital metastases are uncommon. it represents 1-13% of all orbital tumors, with a prevalence in cancer patients estimated to range from 2 to 5% [1].

The incidence of metastatic orbital tumors is highly variable, according to geographical area and race. It has been more increased in recent years due to the improvement of the median survival of cancer patients and advances in diagnostic imaging [2]. In addition, medical literature on orbital metastases has played an important role in raising awareness among health professionals responsible for managing these patients, leading to more vigilant surveillance [3].

To date multiple cases of orbital metastases have been described in the literature. Breast, prostate, liver, and lung

cancer are the most common primary cancers that metastasize to the orbit [3]. Although nasopharyngeal carcinomas are characterized by early distant metastases and lymph node dissemination, orbital metastases remain a rare event [4]. To our knowledge, there are only a few reports of pediatric nasopharyngeal carcinoma (NPC) metastatic to the orbit.

In this case report, we illustrate a new case of a pediatric nasopharyngeal carcinoma with orbital metastasis and report the clinicopathological features and therapeutics interventions, as well as the outcomes of this aggressive disease.

II. CASE REPORT

We report the case of a 10-year-old female patient of Moroccan nationality, without notable pathological history, admitted for a 02 months history of a painful left lateral cervical mass, associated with a mild homolateral hearing loss, tinnitus, and otorrhea. The examination objectified a trismus, a fairly good oral condition, reduced light of the oropharynx and the presence of painful bilateral cervical lymphadenopathy.

Nasofibroscope has shown a process of the posterior wall of the cavum, multiple biopsies were done. The histopathological findings showed a histological aspect of undifferentiated carcinoma of nasopharyngeal type: UCNT of cavum (Fig.1).

Cervical-facial Computed tomography (CT) showed a tissue process of the supra-posterior wall, left anterior wall, and left lateral wall of the cavum measuring 45 x 39 x 38 mm, extending to the base of the skull, and the left temporal cerebral parenchyma through the temporal fossa, it also invades the left sphenoidal sinus. Below: it extends to the oropharynx. with bilateral cervical and retropharyngeal lymph nodes metastases (Fig. 2).

The patient received neoadjuvant chemotherapy based on bleomycin, epirubicin, and cisplatin (BEC) protocol with a low radiological response after the third cure. Therefore, the management was completed with concomitant chemoradiotherapy at a total dose of 70 Gy/35 fractions to nasopharynx and lymphadenopathy, microscopic extent (63Gy), and enlarged neck lymph nodes (56 Gy) with weekly cisplatin 50mg/m².

Three months after completing the treatment, bilateral painful palpable masses in the medial canthal area were noted associated with mild proptosis of the left eye, redness, and decreased visual acuity, rapidly evolving. A Cervical facial scan was undergone that revealed bilateral tissue formations of the internal canthus, with hypodense and enhanced wall after contrast, related to secondary locations. The cerebral scan showed a tissue formation of the frontal bone, enhanced after.

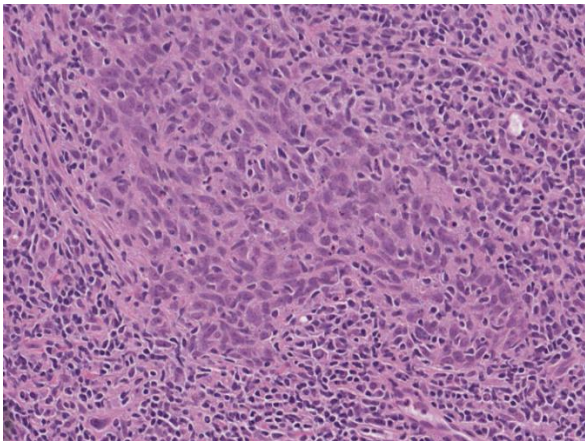


Figure 1. Microphotography showing a syncytial arrangement of carcinomatous cells. Nucleoli are prominent and the stroma is made of an abundant lymphoid infiltrate. HE, 400X



Figure 2 : Axial (A) computed tomography slide shows the primary nasopharyngeal carcinoma with its extensions, and coronal slide (B) demonstrating bilateral cervical lymph nodes metastases.

Contrast without periosteal reaction, related to a secondary location (Fig. 3).

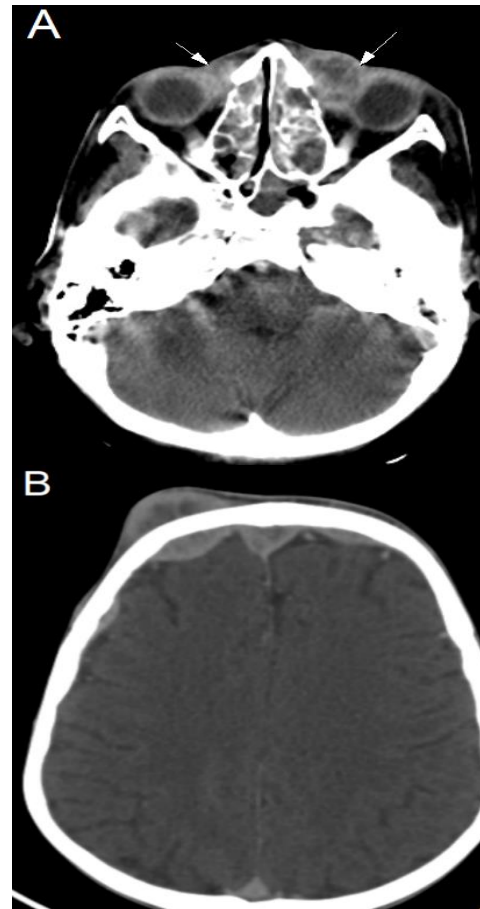


Figure 3 : A: Axial computed tomography slide shows bilateral tissue formations of the internal canthus related to secondary locations.
B : Axial computed tomography slide shows: metastasis of the frontal bone.

Although the patient received two cycles of palliative chemotherapy, the evolution was marked by a progression of orbital metastases (Fig. 4) and died 2 months after the diagnosis of orbital metastases.

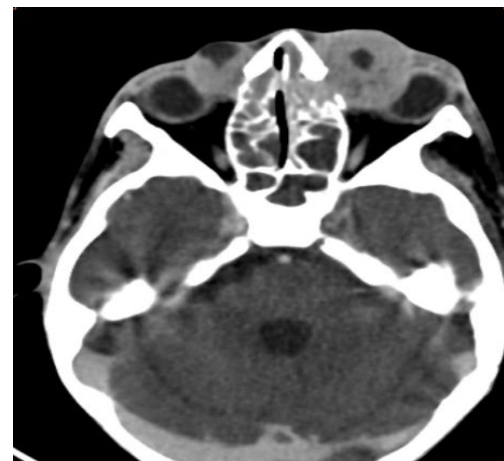


Figure 4 : A: Axial computed tomography slide showing the progression of orbital metastases.

III. DISCUSSION

The incidence of nasopharyngeal carcinoma (NPC) is highly variable worldwide, North Africa is considered as an intermediate risk area where the incidence range from 1 to 10/100,000 person-years. [5] Of all childhood cancers, Nasopharyngeal carcinomas (NPC) represent less than 1%, age distribution is different in endemic regions. only 1 to 2% of all NPC in China occur in children under 16 year versus 10–18% in Mediterranean basin and in Africa. [6]

Of all head and neck cancers, NPC has a highest propensity of early distant metastases which the frequency increase in advanced disease and in recurrent tumors, also and especially in lymph node involvement of the initial tumor, which is consistent with our patient [3-7]. These metastases are present at the time of diagnosis in 10% of cases [8].

Orbital metastases are uncommon, it represents 1-13% of all orbital tumors, with a prevalence in cancer patients estimated to range from 2 to 5% [1]. In addition, Orbital metastasis in children is even more rare. Sarcomas and embryonal tumors of neural origin like neuroblastoma present the most common primary tumors of these metastases in pediatric population [1-2].

In adults, The main primary tumors with orbital metastases are breast with 48%–53% of cases, then lung, and prostate [1]. Although NPC present frequently lymph node spread, orbital metastases remains rare [3]. However, a study from southern China reported that nasopharyngeal carcinoma was the most common primary cancer that metastasized to the orbit with 30.34% of all cases [9].

On the other hand, depending to the volume of the primary tumor, orbital involvement may occur by direct extension through different pathways. The most frequent is by extension to the pterygopalatine fossa and the infratemporal fossa, then to the lower orbital fissure whose most posterior part responds to the orbital apex. [3-7-10]

The clinical presentation of these orbital metastases are rich and not specific [2-5]. In a study published by Char. et al. [11] diplopia was reported in 48% of cases followed by proptosis (26%), pain (19%), decreased vision (16%), and ptosis (10%). In another study, limited ocular motility (54%), proptosis (50%), and palpable mass (43%) were the most frequent clinical findings [12]. It may also uncloud edema or periorbital swelling. Sometimes it can be associated with damage to a cranial nerve [7].

When orbital or ocular metastasis is suspected, A complete history, a detailed ophthalmological examination and a general physical assessment must be established. In patients with no known history of cancer, it is necessary to refer them to an oncologist, in order to undergo a simultaneous general extension assessment in search of the primary tumor. [1-2]

The radiological study is essential in the diagnostic assessment of the disease and the evaluation of local bone and extra-bone extension. Computed tomography (CT) and MRI are the principal means of evaluating suspected orbital lesions. Even though CT is usually the first choice in evaluating the orbit, MRI provides the best resolution of orbital soft tissues. CT may be more appropriate in patients with suspected bone involvement. [1-2-7] Metastatic lesions are most common in the anterior part of the orbit, These lesions are marked by their local aggressiveness reflected by extraocular muscles and bone invasion [1].

After an adequate radiological assessment the histological evidence is not necessary in all cases Indeed, biopsy rarely needs to be performed to confirm metastasis In patients with widespread disease. Sometimes biopsy may be needed for therapeutic decisions. Like performing new targeted therapy by providing molecular study of the tumor, This may be of interest in newly metastatic breast cancer, especially if ocular biopsy is found to be more accessible [1]. At the end of the extension assessment, we considered the orbital involvement of our patient as metastasis in front of its bilaterality as well as the presence of another metastasis of the frontal bone.

Therapeutic management takes into account the patient's performance status, life expectancy and therapeutic effects, The treatment of orbital metastases is aimed at palliative in order to improve patient's quality of life and preserve the functional prognosis [2-3-7] It may include radiotherapy and chemotherapy [2]. NPC is known for its radiosensitivity, so radiotherapy is the main treatment for orbital metastasis from NPC, but chemotherapy also retains its usefulness in some patients [2-3]

Prognosis is generally poor and depends on the primary cancer, In addition Orbital metastases often occur in an advanced stage of disease and in a context of multiple metastases. The overall median survival after orbital diagnosis is 15 months [3,9].

IV. CONCLUSION

Orbital metastasis of nasopharyngeal carcinoma is a rare event in pediatric population. Clinical manifestations of orbital metastases include rapid onset of orbital symptoms like palpable orbital mass with mass effect, diplopia and decrease vision. MRI and CT scan has an essential role for the diagnosis, the treatment is based on palliative radiotherapy and chemotherapy in order to improve patient's quality of life. Although recent therapeutic advances, the prognosis of these patients remains poor.

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