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Periorbital Cellulitis Revealing a Diffuse Large B Cell Lymphoma of the Ethmoid Sinus: A Case Report

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Case Report

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Abstract: Diffuse large B cell lymphoma (DLBCL) of the ethmoid sinus is very rare entity. The presenting symptoms are usually similar to those of benign inflammatory diseases. We report a case of a 50-year-old male patient who presented with acute onset progressive proptosis right periorbital swelling, redness and treated initially as periorbital cellulitis secondary to right maxillary and ethmoidal fungal sinusitis, but proven to have be a diffuse large B-cell lymphoma (DLBCL) of the ethmoid sinus. HIV serology was negative. The patient was referred to the department of oncology where chemotherapeutic treatment for the lymphoma. We emphasize the importance of considering primary lymphoma of the ethmoid sinus in the differential diagnosis of periorbital cellulitis and refractory rhinosinusitis, to help in early diagnosis, which can improve prognosis.

Keywords: Diffuse large B cell lymphoma; Ethmoid; Periorbital cellulitis, Sinusitis.

INTRODUCTION

Sinonasal lymphoma is a rare clinical entity, representing only 1.5% of all lymphomas and 5–8% of extranodal lymphomas of the head and neck [1]. Primary lymphoma of the ethmoid sinus is very rare. The presenting symptoms are usually similar to those of benign inflammatory diseases, making the diagnosis of sinonasal Lymphoma difficult .

Orbital cellulitis is defined as a serious infection that involves the muscle and fat located within the orbit; it can be caused by trauma, upper-respiratory infection, and sinus infection. Although it can occur at any age, it is more common in the paediatric population.

We describe a case of patient in whom initially a diagnosis of periorbital cellulitis was made, but that later proved to be a diffuse large B-cell lymphoma (DLBCL) of the ethmoid sinus. We report the clinical presentation and review the literature, on to alert physicians to this rare condition.

CASE REPORT

A 50-year-old male patient was referred in our Otolaryngology Emergency Department, by ophthalmology with a diagnosis of right sided orbital cellulitis secondary to sinusitis. He was complaining of acute onset progressive proptosis right periorbital swelling, redness, and pain for 20 days, associated to purulent, headache and fever. His medical history was non-contributory.

On examination the patient was febrile with temperature 38°C. He had right periorbital swelling and redness, with inferior displacement and proptosis of the right globe (Figure-1). Visual acuity was conserved, but movement of extraocular muscle was limited. Nasal endoscopy showed purulent rhinorrhea from middle meatus, without visualisation of any tumor mass.

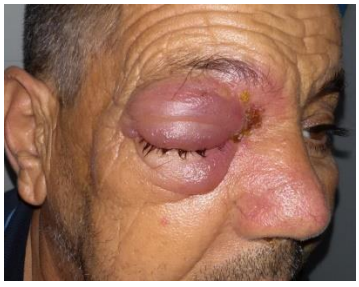

Fig-1: Periorbital swelling and redness, with inferior displacement and proptosis of the right globe

Sinonasal CT scan objective a total opacification of maxillary and ethmoid sinus, with oedema and swelling of periorbital soft tissue. There was proptosis grade 2, thickening of rectus medial muscle and without identification of any intraorbital collection (Figure 2).

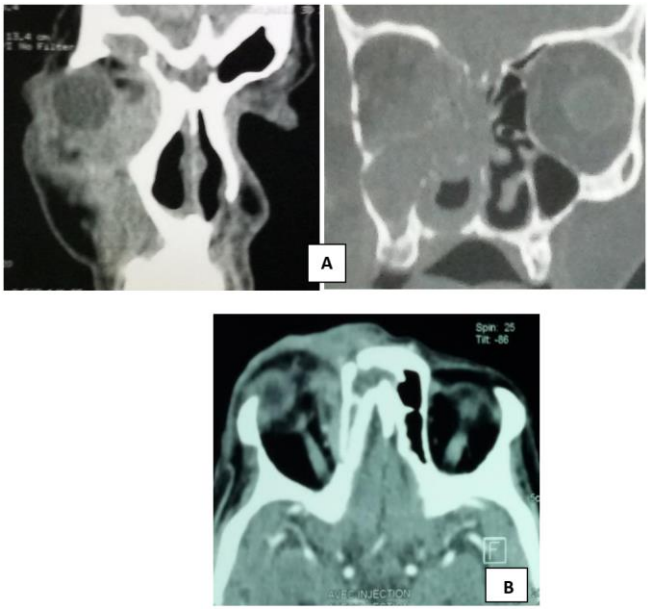

Fig-2: (A) A coronal CT image: Right proptosis, thickening of periorbital tissu and total opacification of ethmoidal and maxillary sinus, with lyse of internal wall of orbit, (B) An axial CT image: Thickening of rectus medial muscle, without intraorbital collection

Laboratory analysis revealed inflammatory syndrome manifested by leucocytosis with C-reactive protein and erythrocyte sedimentation rate were markedly elevated.

The patient was initially diagnosed with periorbital cellulitis secondary to right maxillary and ethmoidal fungal sinusitis and treated with intravenous antibiotic (Cefotaxime and Metronidazole), but without any improvement.

We performed functional endoscopic sinus surgery under general anaesthesia. Middle meatal antrostomy and anterior and posterior ethmoidectomy were done, and tissue was sent for histology. After the surgery, there was no favourable response, orbital symptoms did not improve. Serology HIV was negative.

Histopathological investigation revealed a dense cellular infiltration of lymphoid appearance. The immunohistochemical study phenotyping was negative for markers of T lymphoid cells (AE1 / 3, CD3 and CD56) and found the appearance of a non-Hodgkin lymphoma large B cell phenotype positive to antibodies bcl2 and Ki67**.**

The extension assessment was carried out to the patient, who did not reveal other lymphoma localization. The patient was sent to oncology department, where he received chemotherapy treatment, which consisted on R-CHOP regimen. The evolution is satisfactory after a decline 1 year.

DISCUSSION

Malignant lymphoma of the nasal and paranasal cavities is very rare. In large study of nasal and paranasal sinus malignancy demonstrated that there were no cases of malignant lymphoma of the 220 patients [2].

Hodgkin's lymphoma rarely arises outside lymph nodes, whereas more than 60% of non-Hodgkin lymphoma (NHL) of the head and neck occurs in extranodal sites including paranasal sinuses, nasal cavity, oral cavity, oropharynx, and laryngopharynx [3].

Subtypes of non-Hodgkin's lymphoma presenting in the sinonasal tract include B-cell lymphomas, extranodal natural killer (NK)/T-cell lymphoma (ENKL), nasal type, and other rarer entities [4]

Primary Lymphomas may involve the soft tissues of the external nose, the nasal mucosa, and the paranasal sinuses. Peripheral T cell lymphoma and NK/T cell lymphoma mostly occurred in the nasal cavity, whereas sinus involvement without nasal disease is common in B-cell lymphoma [2]. In the most cases it occurs in the maxillary sinuses, followed by the ethmoid sinuses and the nasal cavity [1].

The prevalence of diffuse large B-cell lymphoma is higher in Caucasian populations, while in Asia and South America nasal NK/T lymphoma predominate [5]. It incidence increases with HIV infection [4].

Clinical manifestation of ethmoidal DLBCL is not specific. Mass effect and progressive mucociliary dysfunction often lead to symptoms consistent with chronic inflammatory sinus disease, including unilateral nasal obstruction, rhinorrhea, pain, recurrent epistaxis and facial oedema. B symptoms (fever > 38º C, nocturnal sweating, and 10% weight loss in six months) occur in 40% to 45% of the cases, and even more frequently in aggressive disease [3].

Orbital cellulitis as the primary clinical appearance of a lymphoma has been reported in the literature before. Charton *et al*., have reported three cases with periorbital involvement; at first they were started on antibiotics until NKTCL diagnosis was confirmed by biopsy [6]

CT and MRI usually show opacification of the sinus with or without sinonasal mass, bone destruction, and invasion of adjacent structures. however, FLourine-18 fluorodeoxyglucose positron emission tomography computerized tomography (18-FDG PET-CT) has better sensitivity [5].

The absence of specificity of clinical and radiological signs can lead to delayed and mis diagnosis and to increase consequently morbidity and mortality.

Repeated and deeper biopsy procedures are recommended to obtain adequate tissue for diagnosis of sinonasal lymphoma. In sometimes surgical management by functional sinus endoscopic surgery endoscopy or Caldwell-Luc techniques are useful for harvesting adequate amount deep tissue for pathological examination [7].

Treatment is exclusively medical. The majority of patients with B-cell lymphoma underwent chemotherapy alone [8]. Chemotherapeutic regimens included CHOP-R and R-ICE; specific choice of chemotherapy was performed on a case-by-case basis by oncologic specialists. The major prognostic factors are delay of diagnosis, initial symptoms, Ann Arbor stage, IPI score and histologic subtype.

CONCLUSION

Large B cell lymphoma is rare entity of malignant disease of the ethmoid sinus that can be easily misdiagnosed as other more common inflammatory processes. Rhinologists should be alert to this aggressive diagnosis and consider it as one of the differential diagnoses of any atypical presentation and non-responsive to conventional treatments of the rhinosinusitis. Early diagnosis and referral to an oncologist could improve the prognosis.

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