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REPORT OF TWO CASES OF MUCOSAL MELANOMA IN THE NASAL CAVITY

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Abstract: Description of the case: Female patients between the eighth and ninth decade of life, with a shared history of controlled arterial hypertension, the second of them with a history of heavy smoking and occupational exposure to the sun; with long-standing clinical pictures, consisting of nasal obstruction and recurrent epistaxis. Clinical findings: on physical examination, deviation of the nasal septum to the opposite side of the lesion, stigmata of bleeding and hypertrophy of the turbinates were observed in both patients, and in extension images evidence of a mass in the nasal cavity with involvement of the maxillary sinus. Treatment and results: both patients required surgical intervention in order to resect the lesion, case #1 was taken to multispecialty surgery and case #2 to surgery solely by otorhinolaryngology, after surgery and currently, both patients are in adjuvant management with radiotherapy. Clinical relevance: mucosal melanoma is a rare but very aggressive form of melanoma that affects the head and neck; Due to its insidious presentation, its diagnosis is usually late, so it is important to know this entity in order to obtain an early diagnosis and timely treatment in order to avoid the spread of its morbidity and mortality.
Keywords: nasal cavity, paranasal sinuses, melanoma, pathology, surgery, adjuvant therapy

INTRODUCTION

Mucosal melanomas are primary malignant tumors that originate from melanocytes that are found spread throughout the body mucosa, the most common being the nasal cavity and maxillary sinuses. Within the nasal cavity, its most frequent location is close to the middle and inferior turbinate, and at the level of the maxillary sinuses, they tend to compromise the maxillary sinus with a greater incidence given its proximity to the nasal cavity.

It is a multifactorial entity, of which predisposing factors such as smoking, exposure to aromatic amines, exposure to ultraviolet radiation, black race, and aging are known; consequently, the incidence of metabolic processes corresponding to the excess of free radicals, epigenetic mutations and genetic predisposition is known.

In terms of epidemiology, it usually occurs equally in men and women, although initially it was believed that it predominated in men due to their occupational predisposing factor for exposure to UV radiation; mainly between the 6th and 7th decade of life and it is a rare form of presentation of the entity; Approximately 20% of all melanomas occur in the head and neck and only 6.3% correspond to the mucosa of this area, resulting in an incidence of 2.2-2.6 cases per 1 million inhabitants. Likewise, the incidence in the general population is only between 0.7-3.8% and is reduced to 0.7-1% in the Caucasian population. Its predominance in the nasal cavity and paranasal sinuses is given, given the high melanocyte population compared to the density of melanocytes in other mucosa.

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The evolution of the clinical picture is usually slow and insidious, which is why patients usually debut in advanced stages of the disease. The most common presenting symptoms are progressive unilateral nasal obstruction over time and epistaxis. Similarly, other manifestations can be bloody spots when blowing the nose, rhinorrhea, local pain, and even epiphora when the melanoma invades the inferior meatus and lacrimal duct.

Computed axial tomography of the facial bones and magnetic resonance imaging are key tools for diagnosis and staging, usually MRI includes T1, T1 post gadolinium, and T2 sequences. However, malignant melanomas

are characterized by heterogeneous contrast enhancement on T1 and T2, which makes diagnosis more difficult. MRI is also the best tool to identify anatomical structures such as the skull and the brain. Finally, positron emission tomography (PET) is a complementary test that could be performed to stratify the tumor, evaluating whether there are distal metastases and to guide the therapeutic approach.

Currently, surgery with or without adjuvant radiotherapy is the treatment of choice and even now, the role of chemotherapy in treating distant metastases is not clear.

With this case report, it was intended and was achieved to elucidate mucosal melanoma as an entity difficult to diagnose and consequently difficult to manage when diagnosed late, in order to collaborate this way in raising awareness of the disease, its timely diagnosis and treatment. early to avoid the high associated morbidity and mortality.

CASE DESCRIPTION:

For the clinical cases presented below, the informed consent signed by the patients and/or their direct guardian is obtained.

CASE 1:

Female patient, 84 years old, with a history of arterial hypertension under pharmacological management with losartan, who consults for left nasal obstruction of one year of evolution, accompanied for the last 3 months by recurrent ipsilateral epistaxis and who brings CT of paranasal sinuses with evidence of probable left antrochoanal polyp versus inverted papilloma, hypertrophy of middle and inferior nasal turbinates and septal deviation. On physical examination on admission, anterior rhinoscopy showed nasal septum deviated to the left, with right nasal spur, mass obstructing the lumen of the left nasal corridor, bilateral middle and inferior turbinate hypertrophy and stigma of bleeding in both nostrils. On admission to the institution with pathology that evidenced the presence of neoplastic cells, with immunohistochemistry with positivity for SOX-1 and negativity for epithelial markers, CD56 and desmin that makes it histologically and immunophenotypically compatible with tumor melanoma; Additionally, brain MRI showed primary pathology of the left nasal cavities with bony distribution and involvement of the left maxillary sinus and ipsilateral orbital wall, signs of pansinusopathy due to occlusion of the osteomeatal complexes and fazekas 1 microangiopathic lesions. She was evaluated together with neurosurgery, with whom it was considered to resect the skull base lesion via endonasal extended by multispecialty, due to probable compromise of the dura mater and requiring CSF fistula closure due to expected coverage defect. Finally, taken to surgery with the finding of a mass completely obstructing the left nostril, with septal deviation to the right, tumor infiltration in posterior septum and lateral wall of the nostril, left middle meatus, left middle and inferior turbinates, involvement of left choana and Eustachian tube, infiltration of the mucosa of the maxillary antrum, infiltration of the left sphenoid mucosa, exposure of the dura mater with tumor infiltration, achieving almost total resection of the lesion. Patient with satisfactory postoperative intrahospital, discharged without complications and on an outpatient basis began cycles of radiotherapy, clinical oncology ruled out the start of chemotherapy.

CASE 2:

Female patient, of African descent, originally from the department of Cauca-Colombia, 77 years old, with a history of pharmacologically controlled arterial hypertension, resolved glaucoma and heavy

smoking that ceased 20 years ago. In addition, he refers to an occupational history of exposure to ultraviolet radiation due to having worked in agriculture for most of his life.

He consulted for a clinical picture of one year of evolution, consisting of obstructive nasal symptoms and recurrent moderate epistaxis rapidly progressive to severe in the clinical course, with physical examination findings of nasal septum deviated to the left, hypertrophy of inferior turbinate, erythema and edema in nasal corridor, mass in right nostril obstructing the lumen, compatible with polyp. She had a simple and contrasted paranasal sinus MRI at admission, which reported an expansive lesion in the right nostril with involvement of the right maxillary sinus, the appearance suggests an antrochoanal polyp without being able to rule out infiltrative involvement, therefore, it was considered to be taken to resection of the lesion with antrostomy. During surgery, a hyperpigmented and friable exophytic mass was found involving the right nostril and hyperpigmented infiltration of the mucosa. A biopsy of the lesions was performed, with positive intraoperative pathology for invasive melanoma and molecular biology tests showed BRAF gene (Exon 11 and 15): not mutated. Immediate postoperative discharge and outpatient management with adjuvant radiotherapy was started.

DISCUSSION

Mucosal melanoma is a multifactorial disease, which usually appears late in life and presents aggressively. It is believed that predisposing factors such as exposure to UV radiation and smoking have a high influence in its appearance.

Because of its slow and insidious onset, it is usually diagnosed in advanced stages of the disease, with few specific symptoms such as nasal obstruction and long-standing epistaxis.

This makes it a disease with poor prognosis due to its high tumor aggressiveness and late detection. MRI is the diagnostic imaging since it allows visualization of the anatomical structures and the treatment of choice is tumor resection with or without adjuvant radiotherapy.

In the case of our two patients, the natural history is compatible with their clinical evolution, as well as the late presentation, both with long-standing clinical symptoms consisting of nasal obstruction and recurrent epistaxis, with predisposing risk factors and compatible imaging and intraoperative findings.

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