Case reports

Olfactory neuroblastoma

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Neuroblastoma olfatorio

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The authors declare there is no conflict of interest.

Abstract
Case presentation. A 36-year-old man, with a three-month history of right palpebral swelling, accompanied by epiphora, photophobia and in the right eye, he only presented perception of light stimulus. Radiological studies identified changes in the density of the paranasal sinuses, mainly on the right, and occupation at the level of the right maxillary sinus. **Treatment.** Complete surgical excision of the lesion was required. According to the immunohistochemical findings, an olfactory neuroblastoma of the right nasal region was diagnosed. Due to the severity and rapid progression of the disease, were included post-operative neurocritical care, mechanical ventilation with tracheostomy and aminergic support. **Outcome.** Evolution was unfavorable, the patient died after 18 days of hospital stay.

Keywords
Olfactory neuroblastoma, neuroepithelioma, olfactory mucosa.

Resumen
Presentación del caso. Hombre de 36 años, con cuadro de inflamación palpebral derecha de tres meses de evolución, acompañado de epífora, fotofobia y en el ojo derecho únicamente presentaba percepción de estímulos luminosos. Los estudios radiológicos identificaron cambios en la densidad de los senos paranasales a predominio derecho y ocupación a nivel del seno maxilar derecho. **Intervención terapéutica.** Se realiza exéresis quirúrgica completa de la lesión. De acuerdo con los hallazgos inmunohistoquímicos se diagnosticó un neuroblastoma olfatorio de la región nasal derecha. Debido a la severidad y la rápida progresión de la enfermedad. Se incluyeron cuidados neurocríticos posteriores a la intervención, ventilación mecánica con traqueostomía y soporte aminérgico. **Evolución clínica.** Su evolución no fue favorable; el paciente falleció después de 18 días de estancia hospitalaria.

Palabras clave
Neuroblastoma olfatorio, epitelio olfatorio, cavidad nasal.

Introduction

Olfactory neuroblastoma, also known as esthesioneuroblastoma[1], is a tumor derived from the basal layer of the olfactory epithelium[2]. It is presented as a soft tissue mass in the upper portion of the nasal cavity involving the anterior and middle ethmoid air cells on one side and it is extended through the cribriform plate in the anterior cranial fossa[1,2].

In 1924, an olfactory nerve tumor was described by Berger, Luc, and Richard that seemed a histologically retinoblastoma, and it was called olfactory neuroblastoma[3]. Two years later, a second intranasal neurogenic tumor was described by Berger and Coutard[4] that differs somewhat in histological pattern and it was called esthesioneurocytoma.This tumor was reported by Seaman and Schall and Lineback[5] in the American literature for the first time in 1951.

Olfactory neuroblastoma is rare and represents less than 3 % of intranasal neoplasms[1,2]. According to epidemiological evolution, its age distribution is bimodal with a peak in adult patients in the second decade of life and another in the fifth and sixth ones, without a predilection for recognized gender[1,6].

Case report

A 36-year-old man, originally from the central area of El Salvador, with no relevant medical, surgical or family history, who went to the General Hospital of the Salvadoran Social Security Institute on April 2020, due to a picture of right palpebral inflammation, accompanied by epiphora and photophobia in the right eye of 3 months of evolution with management of apparent allergic conjunctivitis in a peripheral unit, without clinical improvement.

Afebrile, alert, oriented, with heart rate of 66 beats per minute, respiratory rate of 14 per minute and blood pressure of 120/70 mmHg was found on physical examination. In the craniofacial examination, the right palpebral edema was identified with mild pain on palpation, accompanied by blushing, heat and only perception of light stimuli in the right eye was presented. Rhinoscopy showed a polypoid tumor that occupied 70 % of the right nostril. It was located in the upper third of the nostril and it was extended to the nasal septum and the middle turbinate without invading them; the inferior turbinates had no infiltration and lesions of tumor or inflammatory type were presented in the left nostril. Another abnormality was not identified on physical examination.

Laboratory data indicated mild anaemia (haemoglobin 11,6 g/dl), without leukocytosis or neutrophilia, platelets, creatinine and random glucose in normal ranges and C-reactive protein of 1,22 mg/L. Paranasal sinus x-ray in lateral and right Cadwell projections showed asymmetry with loss of differentiation of the nasal turbinates to right predominance and asymmetry of the soft tissues of the frontonasal region (Figure 1).

Other imaging studies of the paranasal sinuses were performed. A mass of soft tissues with infiltration into both maxillary sinuses, medial walls of the orbits, and displacement to the right medial rectus muscle laterally was described by the computerized axial tomography. Brain Magnetic Resonance showed a lesion of infiltrating features to cribriform lamina and frontal bone with extension to right intraconal region. According to Kadish, it was classified as type C1 (Figura 2).

The imaging findings were evaluated by the specialties of otolaryngology and neurosurgery, who analyzed the rapid progress and stage of the nasal tumor, compatible with an olfactory neuroblastoma, with probable diagnoses of rhabdomino-

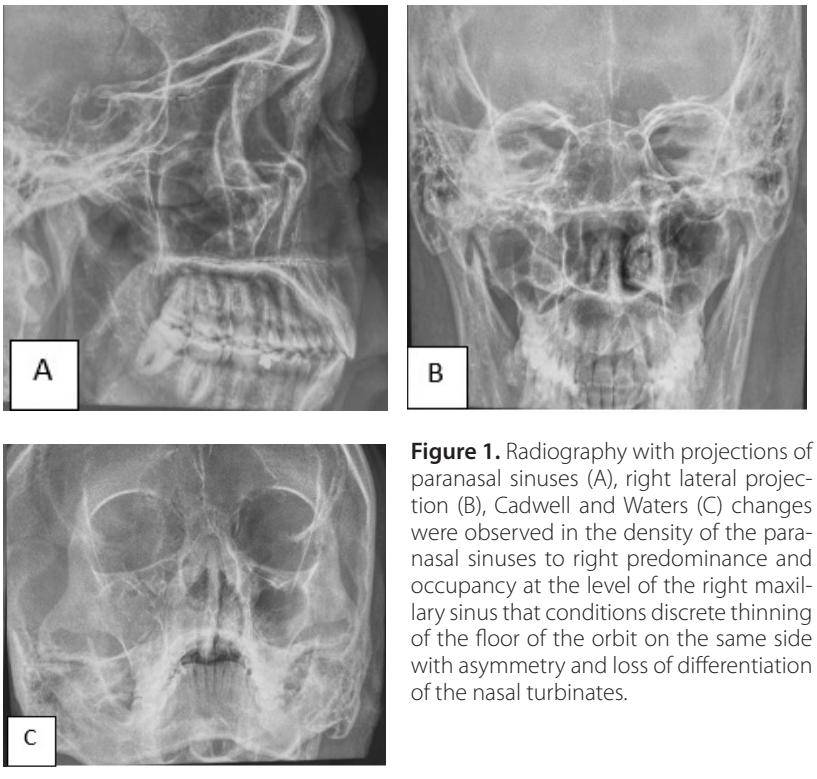

Figure 1. Radiography with projections of paranasal sinuses (A), right lateral projection (B), Cadwell and Waters (C) changes were observed in the density of the paranasal sinuses to right predominance and occupancy at the level of the right maxillary sinus that conditions discrete thinning of the floor of the orbit on the same side with asymmetry and loss of differentiation of the nasal turbinates.

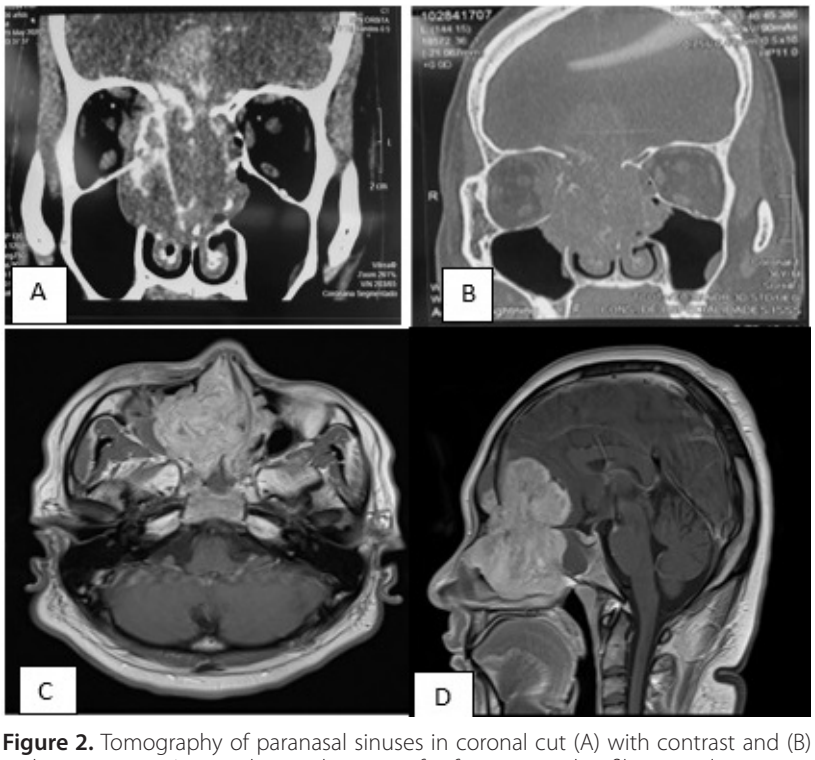

Figure 2. Tomography of paranasal sinuses in coronal cut (A) with contrast and (B) without contrast. It was observed a mass of soft tissues with infiltration, destruction and extension to both maxillary sinuses, medial walls of the orbits and displacement to the right medial rectus muscle laterally. Cerebral magnetic resonance imaging (C) T1 axial cut and (D) sagittal cut with ferromagnetic contrast (gadolinium), which showed an avid enhancement of the lesion, of infiltrating characteristics to cribriform lamina and frontal bone with extension to right intraconal region. It had no local extension to cervical or thoracic nodes (data not evaluable in cuts presented).

sarcoma, nasopharyngeal carcinoma or sinonasal carcinoma.

Therapeutic intervention

The multidisciplinary team decided the early surgical intervention through the nasal hybrid endoscopic approach combined with frontal anterior exeresis. The transsurgical findings were limited to the tumor with soft tissue involvement, intracranial extension and the complete excision of the lesion was achieved, covering both maxillary sinuses, right orbital floor and infiltration to the right intraconal region with its respective optic nerve and the enucleation of the right eyeball was decided. The surgical time of the procedure was seven hours long. The patient required invasive mechanical ventilation and transfer to the intensive care unit.

A histopathological study of samples of 0,3 and 0,7 cm was sent, which it was reported the presence of medium-sized monomorphic cells, with hyperchromatic nuclei, moderate pleomorphism, broad cytoplasm clarified and some eosinophilic, with focal formation of Homer-Wright rosettes (Figure 3).

Clinical evolution

The patient received neurocritical care after the intervention, mechanical ventilation with tracheostomy and aminergic support. The patient died in critical care during hospital stay due to the severity and extent of the injury. According to the parameters of severity and intracranial extension, tumors considered Kadish C type, with intracranial extension, have a mortality rate near 92 % in the trans and postoperative period, even if the complete excision of the lesion is successful[17].

Clinical diagnosis

According to the findings in immunohistochemistry in chromogranin, synaptophysin and CD56 stains, the diagnosis of olfactory neuroblastoma of the right nasal region was reached.

Discussion

Olfactory neuroblastoma usually presents nonspecific symptoms, which makes an early diagnosis difficult[1]. The average delay between the first symptom and diagnosis is six months. In 70 % of cases, bilateral nasal obstruction, epistaxis, hyposmia and headache appear, symptoms that are common in other pathologies, such as allergic polypoid sinus disease or chronic rhinosinusitis[7].

Patients often consult in advanced stages with anosmia and visual disturbances[1], destruction of nearby structures, such as the orbit, brain, facial soft tissues and skin, due to large tumors, which can spread to the intracranial compartment[1].This invasion may be superior in the anterior cranial fossa, laterally in the orbits and through the midline into the contralateral nasal cavity. They can also obstruct the osteomeatal unit of the sinuses, resulting in opacification of the breast with secretions[1]. Locally, signs of invasion may be shown by advanced disease[6].

The importance of the clinical approach, image and early therapeutic diagnosis makes an early multidisciplinary approach essential in future patients[8].

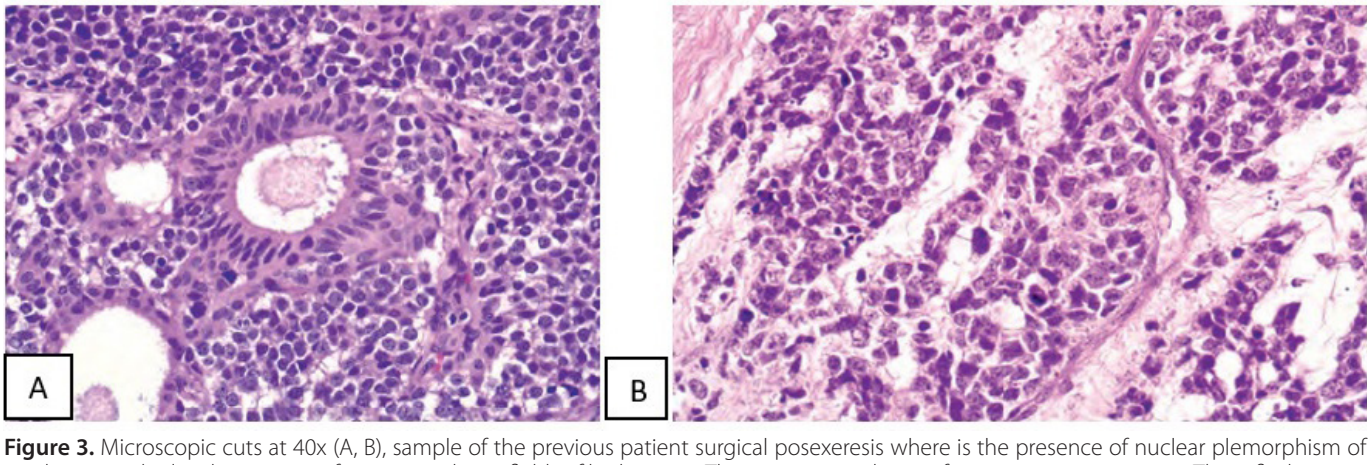

Figure 3. Microscopic cuts at 40x (A, B), sample of the previous patient surgical posexeresis where is the presence of nuclear plemorphism of moderate marked with presence of 20 mitoses by 10 fields of high power. There was no evidence of necrosis on examination. These findings are compatible with olfactory neuroblastoma.

A rhinoscopy[7] can show a mass indistinguishable from other conditions, such as polyposis, chronic sinusitis or other malignancies of the nasal cavity[8].

La Computed axial tomography and magnetic resonance imaging are important to identify the extent, stage of the tumor and surgical approach[1].

PMultiple strategies have been used for the management of this disease, from exclusive surgery, the associations of radiotherapy and chemotherapy, surgery and radiotherapy to the association of the three therapeutic weapons: surgery, radiotherapy and chemotherapy[9-14].

Five-year survival is approximately 70 %[15]. The therapeutic strategy that seems to achieve the lowest rate of local recurrence (10 %)[16] is surgery with craniofacial resection, followed by external radiotherapy, being possible, after local recurrence, therapeutic rescue in 33-50 %[1,17] of cases.

Ethical aspects

The case presented reflects information obtained from clinical records. Neither intervention nor intentional modification of patient data was performed. Patient confidentiality was respected and the data in the publication has been used for academic purposes.

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