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Case Report

Secondary headache as a presenting symptom of sinonasal undifferentiated carcinoma mimicking paranasal mucocele: case report and diagnostic implications

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Background
Secondary headaches can signal serious underlying conditions and require early recognition to guide appropriate diagnostic and therapeutic interventions. The SNOOP10 criteria assist clinicians in identifying red flags suggestive of secondary causes. Sinonasal tumors, such as mucoceles and undifferentiated carcinomas, may present with similar symptoms and radiological features, making differentiation challenging without histological confirmation.

Case Presentation
We report the case of a 32-year-old woman with a one-year history of right-sided pressing periocular headaches, initially responsive to NSAIDs. Over time, the pain intensified, became less responsive to treatment, and was associated with visual impairment, ptosis, and periorbital paresthesia. Imaging revealed a right maxillary paranasal mass initially suspected to be a mucocele. However, biopsy and immunohistochemistry confirmed a diagnosis of sinonasal undifferentiated carcinoma (SNUC). The patient underwent radiotherapy, with partial recovery of ocular motility and ptosis, though visual loss persisted.

Conclusion
This case illustrates the importance of recognizing red flags in headache evaluation and highlights the utility of the SNOOP10 tool in identifying secondary headache disorders. In patients with atypical headache patterns and orbital involvement, early imaging and biopsy are essential for accurate diagnosis and timely management.

Keywords:
Secondary headache
Sinonasal undifferentiated carcinoma
Paranasal mucocele
SNOOP10
Orbital involvement
Cranial neuropathy

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Introduction

Secondary headaches are a manifestation of a serious underlying pathology, the SNOOP10 scale allows them to be identified in order to make precise and timely therapeutic decisions, thus reducing complications, permanent disability and mortality. The growth of tumors in the paranasal sinuses causes varied symptoms due to bone erosion and compression of surrounding structures (including nervous tissue). Mucoceles (benign tumors) are generated by the encystment of viscous mucus produced by the secretory epithelium of the paranasal sinus. In the case of pediatric patients, it is associated with mucoviscidosis (cystic fibrosis). The best treatment option is surgery; on the other hand, sinonasal undifferentiated carcinoma (malignant neoplasm that can be associated with Ebstein Bar virus infection, is highly aggressive and extremely rare) whose best therapeutic approach is multimodal where radiotherapy, chemotherapy and surgery are combined; It has in common with mucocele the anatomical locations, similar initial symptoms and similar radiological appearance in imaging studies. The only way to differentiate them is through biopsy. We present the case of a 32-year-old patient with headache who changed her pain pattern, had little response to drugs and developed compressive optic neuropathy secondary to the presence of a tumor initially classified as maxillary paranasal mucocele.

Case Report

The case of a 32-year-old woman of mixed race, a housewife, is presented. She sought medical attention for experiencing oppressive headaches for the past year, starting as right periocular pain with ipsilateral hemicranial radiation, intensity 5/10 (worsening with physical effort), preceded by visual flashes of light (lasting from minutes to hours). The headaches lasted for 2 hours with a frequency of 10 episodes per month, initially responding to high doses of NSAIDs. Five months before her visit, the intensity of the pain increased to an EVA (Visual Analog Scale) of 8/10 (worsening with ocular movement), and her response to analgesics decreased. She reported decreased visual acuity in her right eye and noticed the presence of blind spots in the visual field of the affected eye. Additionally, she experienced a loss of sensitivity in the infraocular region and episodes of paresthesia resembling electric shocks in the right eye and around it. She was diagnosed with migraine with visual aura and was prescribed treatment with pregabalin 150 mg every 24 hours and dexamethasone 4 mg every 12 hours for 7 days during the pain episodes, which led to partial pain remission with an EVA of 4/10.

She currently seeks consultation for intense retroocular oppressive pain, loss of vision in the affected eye, and ptosis. She denies experiencing nasopharyngeal symptoms (nasal obstruction, rhinorrhea, epistaxis, etc.), denies any craniofacial traumatic history, and has no other comorbidities.

Physical examination

Right eye with ptosis, painful right ophthalmoplegia (Figure 1), absence of pupillary reflexes, amaurosis (involvement of cranial nerves II, III, IV, and VI), mild proptosis, tenderness on palpation in the right periocular and paranasal regions (involvement of the V2 nerve), evidence of mass protrusion in the upper portion of the lateral wall of the right nasal cavity.

SNOOP10 Scale (alarm signs)

Neurological deficit, change in pain pattern, pain worsening with physical activity, progressively increasing headache, ocular pain.

Based on the features described, a secondary headache is suspected.

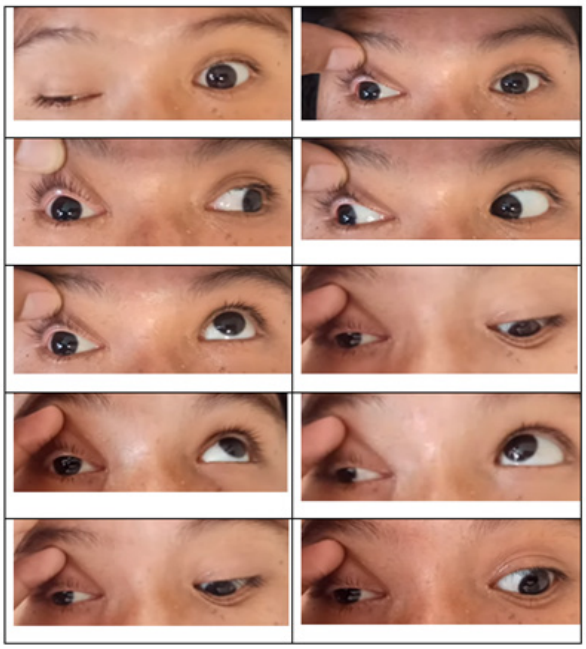

Figure 1. Right ptosis and ophthalmoplegia with partial preservation of abduction (involvement of cranial nerves II, III, IV, and VI).

Computadorizes tomography

Right maxillary paranasal sinus mass of 3.39 x 2.57 x 4.43 cm compressing the right orbital floor and the ocular fissure, deforming the bony architecture, compatible with a mucocele (Figure 2).

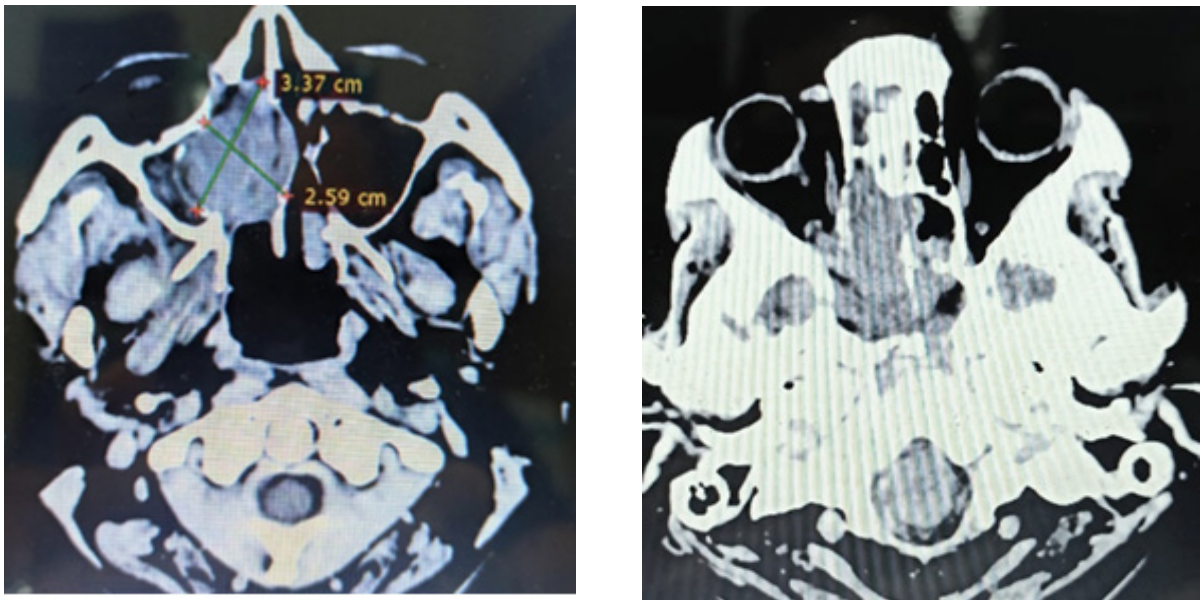

Figure 2. Right maxillary paranasal tumor measuring 3.39 x 2.57 x 4.43 cm, probable mucocele.

Biopsy result

Sinonasal undifferentiated carcinoma

Immunohistochemistry studies

- Cytokeratin OSCAR: Positive
- Cytokeratin 5/6: Positive in isolated cells
- SOX10: Positive
- Actin: Positive in stromal component
- Synaptophysin: Positive in isolated cells
- Epstein-Barr Virus: Negative

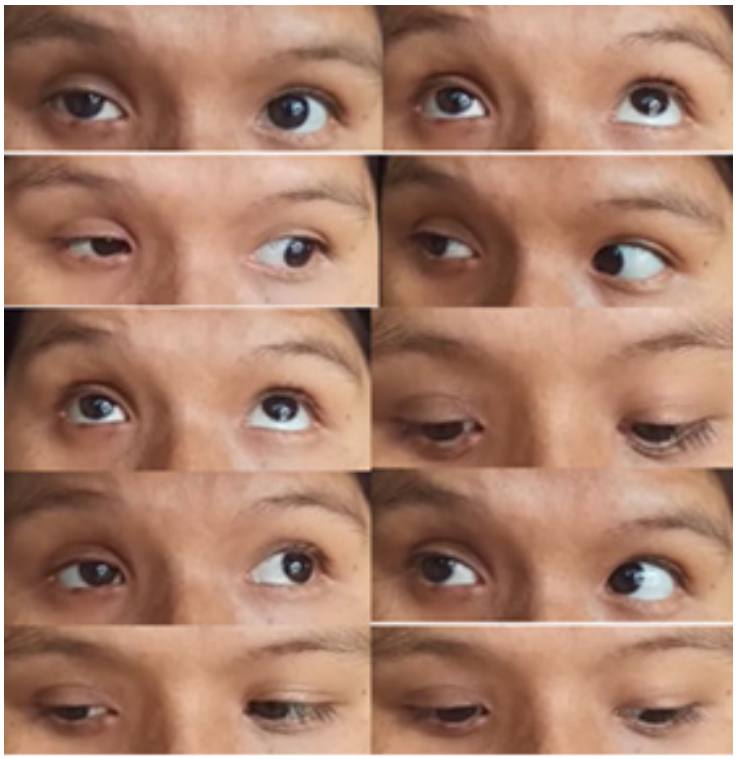

Figure 3. Partial resolution of ophthalmoplegia and ptosis after radiotherapy.

Treatment

The patient received 20 sessions of radiotherapy, followed by chemotherapy, and will be reassessed to determine the need for surgical treatment. After the radiotherapy sessions, there was an improvement in pain reduction, significant recovery of eyelid elevation, and ocular movements. However, visual function was not recovered (Figure 3).

Discussion

Headaches are a common reason for consultation. The scale SNOOP10 (1) is of great importance to guide the anamnesis and physical examination to detect high-risk secondary headaches (2). In this case, after finding some alarm signs (NOP4) – it is not necessary to complete the entire SNOOP10 acronym – a computadorize tomography was requested, which showed the presence of a tumor in the right maxillary paranasal sinus that was categorized as a mucocele due to its location and morphology. Mucoceles typically occur in individuals between 40 to 60 years old; they are rarely seen in pediatric patients. In children, they are associated with cystic fibrosis in up to 60% (3). They can be located in any of the paranasal sinuses (frontal 60%, ethmoidal 30%, maxillary 10%, and sphenoidal 3%) (4). They can be primary or secondary (the main causes are: chronic rhinosinusitis 50.1%, craniofacial trauma 17.4%, or paranasal surgery 19.6% (4,5). In 80% of cases, it erodes the walls of the orbit, and its symptoms are usually headache 42%, maxillofacial pain 28%, proptosis 15%, periorbital pain 12%, and visual disturbances 8% (5,6). The main causes of mortality are cerebrospinal fluid fistulas and meningitis (7). Endoscopic nasal marsupialization is the most recommended surgical technique due to its safety, absence of facial scars, and low recurrence rate (recurrence with open surgery: 25% at 4 years) (4,8).

The tumor biopsy of our patient was reported as sinonasal undifferentiated carcinoma, a malignant neoplasm whose association with Epstein-Barr virus infection is not well established (9,10). It is highly aggressive (at the time of diagnosis, 50% have dural invasion and 30% have orbital invasion) (11). It is extremely rare (3-5% of nasosinusoidal neoplasms), with an age of presentation ranging from 30 to 90 years. It predominates in men (M:F = 2-3:1). Its symptoms are nonspecific (nasal obstruction, epistaxis, craniofacial pain, upper dental pain, headache, proptosis, diplopia, optic neuropathy, decreased visual acuity, paralysis, and paresthesia of cranial nerves). Due to this, it is diagnosed in advanced stages (T4) (10–12) 10 to 30% of patients have cervical lymphatic metastasis (11). Imaging from contrast-enhanced tomography or magnetic resonance imaging reveals non-calcified masses with remodeling of adjacent structures and variable contrast uptake (9,11). It has a median survival of 22.1 months (34.9% of patients survive 5 years, and 31.3% survive 10 years). The best therapeutic options are multimodal, with surgery followed by radiotherapy achieving a relative 5-year survival rate of up to 39.1% (surgery alone 38% or radiotherapy alone 36%) (13). The prognosis worsens if there are cervical metastases, distant metastases, or positive resection margins (14). Recently, induction chemotherapy has been described to achieve chemo-selection and tumor cytoreduction, and if there is a good response, concurrent chemoradiotherapy is favored, which has been associated with better overall survival and a higher rate of organ preservation (15).

Conclusions

The diagnosis of migraine with aura, without a proper case analysis, delayed the diagnostic decision-making, allowing the progression of the disease for a year until the patient was evaluated with the SNOOP10 scale, which helped identify the case as a secondary headache and determined the need for a tomographic study. Since sinonasal undifferentiated carcinoma (SNUC) does not have distinct radiological characteristics, we emphasize the importance of biopsy and immunohistochemical studies prior to any surgical resection of paranasal tumors in order to make precise therapeutic decisions based on each pathology. In the presented case, there is evidence of rapid progression of clinical manifestations: one year of headaches, and in the last 5 months, visual impairment, paralysis of oculomotor nerves, and paresthesia of sensory nerves (V2), which is consistent with the clinical manifestations and rapid progression over time described by the cited authors. This article also provides a description of the characteristics of oppressive headaches, starting periocularly (paranasal and infraorbital). There is no reported SNUC case series in Latin America.

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