Atypical Presentation of a Sinonasal Adenocarcinoma Clinically Mimicking a Mucocele: Case Report

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Abstract
Sinonasal adenocarcinomas are rare, it represent 4 to 20% of nasosinusian carcinomas, the latest WHO 2017 classification distinguishes two types: Intestinal type adenocarcinomas (ITAC) and non-intestinal type, with different prognosis; intestinal adenocarcinomas being more aggressive with a mortality rate around 53%; nasal obstruction, epistaxis, and rhinorrhea are common symptoms, the rhinoscopy found multiple fleshy, bleeding formations in contact, occupying nasal cavities first evokes a mucocele.

We present the case of a 53-year-old patient who reported a rapidly growing soft mass on his nasal pyramid for 10 months, with nasal obstruction, bilateral exophthalmos and blindness of the right eye, on the CT scan of sinuses, we observed a heterogeneous formation, associated with adjacent bone destruction with intracranial extension, directing the diagnosis to a neoplasm sinonasal malignant; A deep biopsy under general anesthesia was made, the histological study concluded to intestinal sinonasal adenocarcinoma; which are rare.

High grade sinonasal adenocarcinomas are locally aggressive tumors often diagnosed at an advanced stage. Their treatment is mainly surgical and the prognosis depends essentially on the clinical stage; therefore the diagnosis should not be delayed.

Our case underlines the importance to always maintain a high clinical index of suspicion of malignancy, in particular in cases of extensive mucocele

Keywords
Adenocarcinoma, Nasal Cavity, Mucocele

Introduction
Carcinomas of the nasal cavity and paranasal sinus are rare, with an annual incidence of less than 1 per 100,000 individuals [1,2]. The latest WHO 2017 classification distinguishes two types of adenocarcinomas: Intestinal type adenocarcinomas (ITAC) and non-intestinal type, with different prognosis implications [3]; intestinal type adenocarcinomas are more aggressive with a mortality rate of around 53% [4], Sinonasal intestinal type adenocarcinomas are clinically aggressive and diagnosed most often at an advanced stage with a mortality of 53% [4]; however metastases are relatively rare, and the prognosis is mainly attributed to clinical stage and recurrences frequency [5]. Sinonasal intestinal type adenocarcinomas makes up only a small percent of all nasal and paranasal sinus malignancies [3], they occur sporadically or are associated with occupational exposure to hardwood and leather dust, nickel, and possibly smoking [6]. The relationship between exposure to wood dust and the development of intestinal-type adenocarcinoma has been established by numerous epidemiological studies [6-8]. The preferential site of sinonasal adenocarcinoma is the ethmoidal sinus [3] and men are 5 times as likely to develop these tumours as women [4]. Symptoms are often a common rhinologic pathology [9] Radiologic examination is based on computed tomography (CT) scan and magnetic resonance imagery (MRI). The most commonly employed treatment has been a combined treatment associating surgery and radiotherapy. We describe a rare case of sinonasal adenocarcinoma which presents clinical atypical mimicking a mucocele.
Figure 1: Prefistulization nasal mass with bilateral exophthalmos.

Figure 2: CT of the paranasal sinuses in the soft tissue window, showing an expansive lesion, with areas of necrosis, bone destruction with bilateral and intracranial orbital dissemination.

Case Presentation

We report the case of a 53-year-old patient, with 10-years history of smoke; reported a rapidly growing soft mass on his nasal pyramid for 10 months, with bilateral exophthalmos, blindness in the right eye and permanent bilateral nasal obstruction, associated with intermittent epistaxis, and anosmia. (Figure 1). The patient subsequently reported insomnia, and intense headache that was resistant to symptomatic medical treatment (paracetamol). The rhinoscopy found multiple fleshy, bleeding formations in contact, occupying nasal cavities; ophthalmologic examination was made objectifying a right optical atrophy and a beginning on the left one. The rest of the examination was without particularities.

CT shows a heterogeneous, expansive formation with hypodense areas in relation to necrosis and soft tissue areas, occupying the entire sinus and nasal cavity with intracranial extension, associated to adjacent bone remodeling and destruction, reducing both orbital cavities, laterally displacing both optic nerves, predominantly right, causing bilateral exophthalmos (Figure 2). Two biopsies carried out were not specific; a third deep biopsy was performed under general anesthesia, which histological examination concluded to a high grade intestinal type adenocarcinoma.

After announcing to the patient the diagnosis and detail-
ing the therapeutic modalities as well as the prognosis of his disease, the patient did not adhere and he refused to continue the treatment.

Discussion

Sinonasal intestinal-type adenocarcinoma is a rare neoplasm, with local aggressively behavior and high local recurrence rates [10]. The literature review showed that malignant neoplasms of the sinonasal tract usually present late; 74.7% of all patients was classed stage III and IV disease at the diagnosis [11-16]. The clinical presentation of this tumor is not specific and can simulate a benign tumour, by the presence of nasal obstruction, and epistaxis, our patient presents in addition, clinical signs which predict a local extension such as exophthalmos and intense headaches. The patient was a 53-year-old man who had age and sex risks for sinonasal neoplasms, but no history of exposure to environmental carcinogens, such as leather or wood dust. After, rhinoscopy; we were more oriented to mucocele. CT scan with intravenous contrast is essential to localize the tumor and defines eventual extensions to adjacent structures; the skull of base, orbit and the deep spaces of the face. Additionally, MRI with intravenous contrast complements CT by identifying neoplastic dissemination in the soft tissues. In our case, the massive local extension on the scanner, challenged us on the diagnosis of mucocele, and we had to repeat the biopsy to obtain a diagnosis. In addition; mucocles are benign cystic tumours, arising at the expense of the paranasal sinus mucosa, lined by non-neoplastic epithelium, and containing usually sterile mucus [17]. Given the key role of histology in guiding treatment, it is important that biopsy is obtained with precision and histologic slides are analyzed by a pathologist with specific expertise [18].

Surgery is the most commonly used treatment modality for SNAC; and obtaining negative surgical oncologic margins is essential for tumor control [5]. The endoscopic surgery is the gold standard for the treatment of sinonasal adenocarcinoma, it conditions the prognosis; and adjuvant radiotherapy is recommended in advanced stages and high grade tumors of malignancy [18], moreover advanced stage at diagnosis is acknowledged to have a strong impact on prognosis and, in particular, on the probability of recurrence [19].

Concerning prognosis, disease-related death of sinonasal adenocarcinoma is typically attributed to local recurrence, which occurs in approximately half of cases [17,20]. Metastases are relatively uncommon in sinonasal adenocarcinomas and are usually quoted as occurring in 20% of cases. Lymph node spread occurs in roughly 10% of cases. This behavior underlies the treatment objective of preventing locoregional recurrence and the importance of early diagnosis to manage this neoplasm.

Our case underlines the importance to always maintain a high clinical index of suspicion of malignancy, in particular in cases of extensive mucocele, in order not to delay the diagnosis.

Conflict of Interest

No conflict of interest.