

Sinonasal undifferentiated carcinoma: A case report

Carcinome indifferencie nasosinusien: À propos d'un cas

Dhambri S, Dhaha M, Tbini M, Kedous S, Jebali S, Touati S, Gritli S

Department of Ear, Nose, throat and Head and Neck Surgery, Salah Azaïz Institute of Oncology, Tunis, Tunisia.

Faculty of Medicine of Tunis, University of Tunis El Manar, Tunisia.

Received: 11 July 2019; Accepted: 04 October 2019; Published online: 20 June 2020

ABSTRACT

Objective: The aim was to insist on the main clinical and histological presentations of Sinonasal undifferentiated carcinoma and to discuss the possible treatment modalities reported in the literature.

Observation: a 52-years old painter sought medical consultation with right epistaxis. and ipsilateral recent swelling. Clinical examination found a right 2cm paranasal swelling arasing the naso-labial fold and extending to the right eyeball and nasal pyramid, hypoesthesia in the right V2 territory and an ipsilateral cervical lymph node. Endoscopy concluded a fungating white formation filling the right nasal cavity. CT scan showed a tissular process in the right nasal cavity extended to the ethmoid. biopsy concluded to a sinonasal undifferentiated carcinoma. MRI showed a tissular process of right nasal cavity and the ethmoïd with hyposignal in T1 and hypersignal in T2 with gadolinium enhancement. Chest CT scan showed a suspicious pulmonary node. The patient was classified T2N1M1. chimiotherapy were administrated associated with a flash irradiation in an analgesic purpose. The patient died after a cataclysmic bleeding.

Conclusion: SNUC is a rare tumor of sinonasal tract. It remains challenging due to its aggressive local behavior, its high propensity to regional and distant metastasis and difficulties in its management.

Key-word: Sinonasal undifferentiated carcinoma, Malignant tumors, CT Scan, MRI, Biopsy, Chemotherapy Radiotherapy.

INTRODUCTION

Malignant tumors of sinonasal tract are rare, representing less than 1% of all cancers and 3% of all upper aerodigestive tract cancers [1]. Sinonasal undifferentiated carcinoma (SNUC) is a relatively rare entity specific to the nasal cavities and paranasal sinus. According to the World health organization, SNUC is defined as a highly aggressive and a distinct carcinoma of uncertain histogenesis composed with pleomorphic tumor cells with frequent necrosis that should be differentiated from other carcinomas or olfactory neuroblastoma. SNUC, usually, presents with non specific symptoms such as nasal obstruction, headache and epistaxis for a short duration [2]. Therefore 70% to 100% of tumors were rated T4 at presentation [3-4]. Since it was first described by Frierson and al in 1986, few cases of SNUC were reported [5]. Thus management of these tumors is still unclear guided by small series with small number of patients [4]. Long term survival rates were deceiving reaching 50% in the best cases [6]. The aim of this case report presentation was to insist on the main clinical and histological presentations of the disease and to discuss the possible treatment modalities reported in the literature.

OBSERVATION:

In June 2014, a 52-years old painter sought medical consultation with unilateral epistaxis. Cluster pain of the right hemi face and ipsilateral recent swelling were also reported. The patient was a heavy smoker. Clinical examination found a right 2 cm paranasal swelling arasing in the naso-labial fold and extending to the right eyeball and nasal pyramid (figure1), an hypoesthesia in the right V2 territory and an ipsilateral 2cm cervical lymph node in the level II.



Figure 1: clinical evolution 2 months after diagnosis showing a massive swelling deforming the face

Corresponding Author: Dhambri Sawsen.

Head and neck surgery Department-Salah Azaiez oncology institute, Boulevard du 9-Avril 1938; 1006 Tunis, Tunisia

Email: sawsendhambri@gmail.com

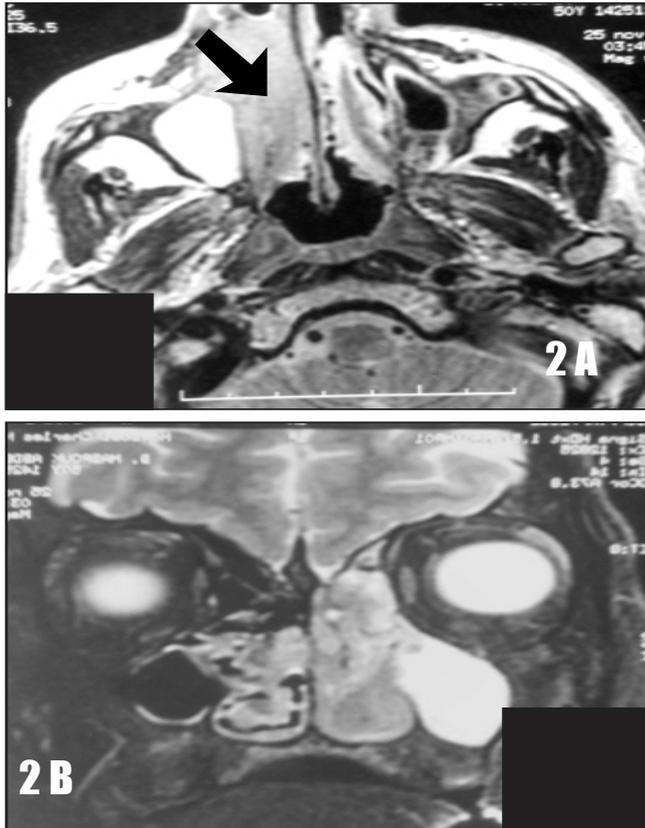


Figure 2A and figure 2B: MRI imaging: T1-gadolinium axial section and T2 coronal section showing a nasal right process infiltrating the ethmoid with retention image in the ipsilateral maxillary sinus.

Endoscopy concluded to a fungating white formation filling the right nasal cavity. CT scan showed a tissular process in the right nasal cavity extended to the ethmoid respecting the floor of the nasal cavity and the palatine vault. biopsy was performed under local anesthesia. Histological examination coupled to immunohistochemistry (IHC) concluded to a sinonasal undifferentiated carcinoma (SNUC) with (PS100-, cytokeratine +, EMA+). MRI showed a tissular process of right nasal cavity and the ethmoid with hyposignal in T1 and hypersignal in T2 with gadolinium enhancement (figures 2A and 2B). Chest CT scan showed a suspicious pulmonary node. The patient was classified T2N1M1. Three cycles of 5FU-CDDP chemotherapy were administrated to the patient. Evolution was marked by rapid growth of the facial swelling complicated with blindness in an interval of 2 months. Flash irradiation in an analgesic purpose where administrated. The patient died after several months after a cataclysmic bleeding.

DISCUSSION:

In 1986, Frierson and al reported the first SNUC series, including 8 patients all with advanced disease with orbital and cranial extension, all were treated with chemo-radiation therapy. Only 3 patients were alive after one year [5]. Since then fewer than 200 cases were reported in the literature [4-6]. Recently, Kuo and al reported a series of 435 patients [7]. This

is to our knowledge the largest cohort ever reported. Worth improvements in the management of SNUC were achieved but many clinical and therapeutic features are still unclear. In addition to its uncertain histogenesis, the etiology of SNUC is to date unknown. The role of cigarettes smoking and previous radiation therapy was not demonstrated.

According to Kuo and al, SNUC was more frequent in males with a sex ratio near to 1.7. The age ranged between 18 and 85 years; half of patients were younger than 55 years [7]. For a short duration, symptoms are non specific with nasal obstruction, epistaxis and headache. Therefore, 70% to 100% were staged T4 at presentation and 10 to 30% had metastatic cervical lymph nodes [3-4]. According to an American review by Caroline C Xu and colleagues, headache and neurologic symptom were present in 45% of cases and epistaxis in 25% [8]. The same review reported that 42.3% of patient had orbital involvement at diagnosis [8]. In our case, cluster trigeminal pain, headache and epistaxis and paranasal swelling were present at the first consultation with a cervical node metastasis. A distant metastasis in the lung was identified and classified the patient as stage IV according to the TNM classification.

Histological examination coupled to immunohistochemistry (IHC) is necessary for the diagnosis. The light microscope features of the SNUC included the presence of a hypercellular proliferation with different growth patterns including trabecular, lobular, sheet-like and organoid patterns [9]. Mitotic index is high with important necrosis and vascular emboli. There is no glandular or squamous differentiation [9]. IHC is necessary for the diagnosis. The immunostaining for the cytokeratine 7, 8 and 19 and the EMA+ [2], like in our case is documented in the literature.

Imaging is fundamental in dealing with malignant tumor of sinonasal tract. While CT scan is very contributive in highlighting bone destruction, MRI is interesting in studying extension to soft part. Tumor limits visible in CT scan are correlated in only 78% of cases to surgical and histological findings [10]. These correlations reach 94% with MRI and 98.4% with gadolinium enhanced MRI [10]. Imaging is necessary in diagnosis, staging and follow-up.

According to Kuo and al, SNUC is difficult to treat for many reasons including, advanced stage at presentation, proximity to critical structures and rarity [7]. No ideal treatment strategy has been systematically evaluated due to the small number of reported cases. Most authors recommend an aggressive therapy to eradicate the diseased tissues [7]. Surgery, when possible, is indicated. It consists usually of mutilating craniofacial resection with maxillectomy, orbital exenteration, and occasionally neurosurgical involvement. Neoadjuvant Chemotherapy associating cyclophosphamide, doxorubicin and vincristin, followed by surgical resection and postoperative or preoperative



radiotherapy is the most common regimen described in the literature [3-6]. According to Mussy and al, these tumors are no longer surgical if one or many of the following structure are involved: cavernous sinus, infratemporal fossa, the brain or the orbital cone bottom [6]. Systematic irradiation of the cervical lymph node compartments for N0 patients is recommended by some authors due to the disease aggressiveness [4-6]. The 5-years survival rates are low. According to Kuo and al, the cumulative 5-years survival rate was 41.5% [7]. Close survival rates were found in other series [9]. Surgery associated to chemotherapy or Chemoradiation therapy is correlated to the best survival rates in univariate and multivariate analysis according to the same authors. In our case, a palliative chemotherapy was administrated to the patient due to the distant pulmonary metastasis.

CONCLUSION:

SNUC is a rare tumor of sinonasal tract of uncertain histogenesis. It remains challenging due to its aggressive local behavior and its high propensity to regional and distant metastasis. Only small retrospective series of patients were reported in the literature making the management of this disease unclear. There is no evidence that aggressive therapy offers better survival. Therefore, a better comprehension of the disease and the search for new modalities for the treatment is necessary.

Compliance with ethical standards

Conflict of interest: The authors stated that there is no conflict of interest.

Funding Statement: The authors received no specific funding for this work.

REFERENCES:

1. Turner JH, Reh DD. Incidence and survival in patients with sinonasal cancer: a historical analysis of population-based data. *Head Neck*. 2012;34(6):877-885.
2. Ejaz A, Wenig BM. Sinonasal undifferentiated carcinoma: clinical and pathologic features and a discussion on classification, cellular differentiation, and differential diagnosis. *Adv Anat Pathol*. 2005;12(3):134-143.
3. Rischin D, Porceddu S, Peters L, Martin J, Corry J, Weih L. Promising results with chemoradiation in patients with sinonasal undifferentiated carcinoma. *Head Neck*. 2004;26(5):435-441.
4. Tanzler ED, Morris CG, Orlando CA, Werning JW, Mendenhall WM. Management of sinonasal undifferentiated carcinoma. *Head Neck*. 2008;30(5):595-599.
5. Frierson HF, Jr., Mills SE, Fechner RE, Taxy JB, Levine PA. Sinonasal undifferentiated carcinoma. An aggressive neoplasm derived from schneiderian epithelium and distinct from olfactory neuroblastoma. *Am J Surg Pathol*. 1986;10(11):771-779.
6. Musy PY, Reibel JF, Levine PA. Sinonasal undifferentiated carcinoma: the search for a better outcome. *Laryngoscope*. 2002;112(8 Pt 1):1450-1455.
7. Kuo P, Manes RP, Schwam ZG, Judson BL. Survival Outcomes for Combined Modality Therapy for Sinonasal Undifferentiated Carcinoma. *Otolaryngol Head Neck Surg*. 2017; 156(1):132-136.
8. Xu CC, Dziegielewski PT, McGaw WT, Seikaly H. Sinonasal undifferentiated carcinoma (SNUC): the Alberta experience and literature review. *J Otolaryngol Head Neck Surg*. 2013; 42:2.
9. Enepekides DJ. Sinonasal undifferentiated carcinoma: an update. *Curr Opin Otolaryngol Head Neck Surg*. 2005; 13(4):222-225.
10. Lund VJ, Howard DJ, Lloyd GA, Cheesman AD. Magnetic resonance imaging of paranasal sinus tumors for craniofacial resection. *Head Neck*. 1989;11(3):279-283.