

Undifferentiated Sinonasal Carcinoma—Case Report

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Abstract: SNUC (undifferentiated sinonasal carcinoma) is a rare tumor. The exact cause of this cancer is not known. Patients usually have signs and symptoms of bleeding from the nose, runny nose, duplicate photos, symptoms of chronic sinus infections and nose, nasal obstruction, or facial pain. We reported a case of seventy year old patient with sinonasal undifferentiated carcinoma in the right maxillary sinus. Six months ago, the patient was conducted operations polyp in the right nasal hall. He had pain in the right side of his face and swelling in the mouth in the area of the upper jaw on the right side. The patient was operated tumor of the right maxillary sinus, with expansion in the ethmoid sinus, nasal cavity and infratemporal space. The neck dissection was done on the same side. Postoperatively, he received radiation therapy and chemotherapy. SNUC is very rare aggressive cancer that requires aggressive therapy. This cancer is considered in need of modified maxillary sinuses, as well as for modified sinus in polyps or after the operation of the same. Despite extensive surgery, radio therapy and implemented chemotherapy, the prognosis of these tumors is very poor.

Key words: Sinonasal, undifferentiated, carcinoma.

1. Introduction

SNUC (Undifferentiated sinonasal carcinoma) is a rare and aggressive type of cancer [1]. This cancer is originating from the epithelium of the paranasal sinuses and nasal cavity [2].

It was first diagnosed in 1987. Years in a patient in an advanced stage of the disease are associated with poor survival. SNUC occurs when cells from the epithelial layer that lines the sinuses form a mass in the sinus by increasing the number of cells metaplasia [2].

The exact cause of this cancer is not known. There are several risk factors that can increase the chances of a SNUC. Risk factors for this cancer are: breathing in certain substances while working, such as dust from the manufacturing of wood, textiles, leather, flour, and metals (including nickel and chromium) [3].

Some patients, states the presence of polyps in the nose or maxillary sinus that are benign in, and that later it detects the presence of carcinoma [4, 5].

SNUC is more common in smokers, and men older than 40 years. Also, the higher probability of this cancer are people who infected with HPV (human papilloma virus), and sometimes it is described EBV (Epstein-barr virus) infections [6, 7].

SNUC is an aggressive cancer, and it is characterized by rapid growth, locally destructive and a tendency of giving distant metastases [5].

Patients usually have signs and symptoms of bleeding or runny nose, duplicate photos, exophthalmos symptoms of chronic sinus infections and nasal obstruction. Usually it occurs due to difficulty breathing through the nose, nasal congestion or facial pain on the same side of the tumor, which is usually mild intensity [2, 5, 8, 9].

2. Case Report

We reported a case of a male patient, seventy years old with a tumor in right maxillary sinus.

The patient complained of pain in the upper jaw on the right side, which started four months ago, immediately after tooth extraction in the upper jaw on the right side (second premolar).

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Patient medical history states that he was conducted operations polyps in the maxillary sinuses twenty years ago. Six months ago he had a stuffy nose and runny hemorrhagic secretions from the nose, when he underwent surgery nasal polyps in the right nasal corridor. He is a smoker already for thirty years, and spends 20 cigarettes a day.

Through clinical examination (Fig. 1), we found palpation hard outgrowth in the area of the right maxilla, above the skin unchanged. Intraoral fornix in the area of the upper jaw on the right side, partly palate presents outgrowth solid consistency, painful on palpation, mucous membranes unchanged. On the neck in the region of the right submandibular region, there is a tumor under the skin surface, oval in shape, measuring about 3×4 cm, firm consistency, moveable in relation to the skin and the limited

vaguely to deeper structures.

Echsonography on neck revealed the presence of reactive lymph glands in the right submandibular gland, size of approximately 35 mm. Chest x-ray was normal. Laboratory findings were normal. Immunological analysis of the patient was negative for EBV.

CT (computerized tomography) (Fig. 2) showed the presence of infiltrative tumor, the size of $65 \text{ mm} \times 40 \text{ mm} \times 45 \text{ mm}$ in the area of the right maxillary sinus with areas of necrosis. Osteolysis was present in the last part of the medial wall of the right maxillary sinus and middle cranial fossa base right wing sphenoidal bones. Tumor spreads to the nasal cavity, right infratemporal space and right ethmoid sinus. Also the present was the content of inflammatory characteristics in the right sphenoidal sinus.

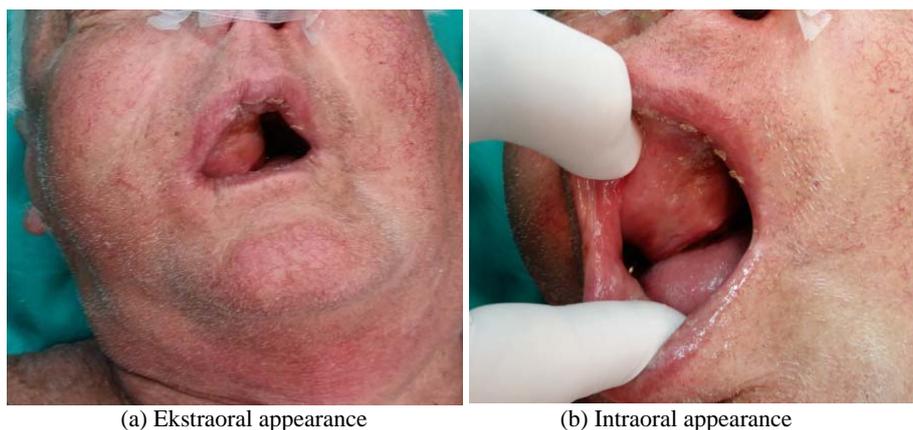


Fig. 1 A patient with a tumor of the maxilla.

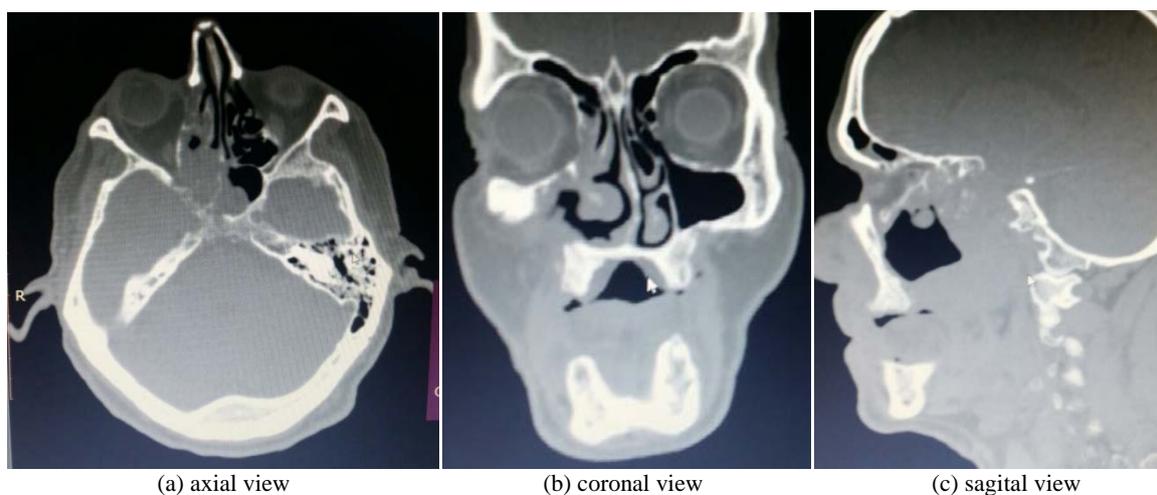


Fig. 2 Computed tomography of the head.

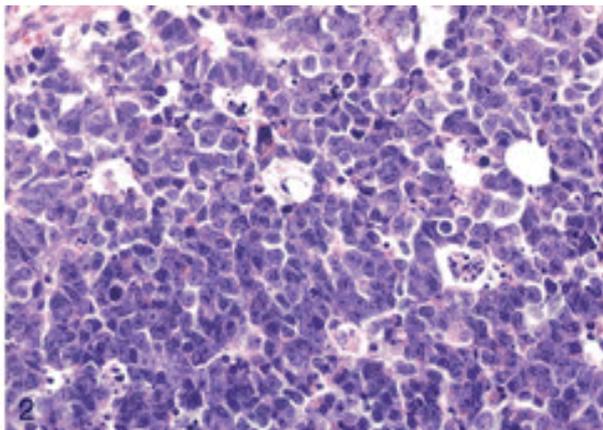


Fig. 3 Sinonasal undifferentiated carcinoma is characterized by malignant cells undifferentiated high grade nuclear morphology (hematoxylin-eosin, original magnification $\times 400$)

On the neck, in the right submandibular region, pathologically enlarged lymph node diameter of 33 mm was present.

The patient underwent surgery under general anesthesia: maxillectomy right, right ethmoidectomy, removal of tumors from the right nasal meatus, right sphenoid sinus, the right infratemporal space and suprahyoid neck dissection on the same side. During the surgical procedure, extempore findings showed that it was a malignant change.

Histology findings (Fig. 3) was diagnosed sinonasal undifferentiated carcinoma. Sinus tumor invades the mucosa, which is constructed of non-standard, large tumor cells arranged in the form of a solid beach and cords, sails are large, present zone of necrosis. Cells are unclear boundaries. Immunohistochemical phenotyping: tumor cells were CK, CK5/6 and CK 19 (+) positive; a CK7, CK8, Nse, CHRA, Syn, NCAM, S100, Melan A, HMB45 melanosomes, LCA, Vimentin and Cd99 negative (-). In preparation of neck dissection, metastatic deposits in the lymph nodes of the submandibular region were found.

3. Discussion

The higher incidence of SNUC in male patients, as it is our patient, is described by other authors and in the literature, which describes the ratio of males and

females from about 2: 1 to 3: 1. SNUC occurs in patients from the second to the ninth decade, and most often in the sixth. Our patient is in the seventh decade of life [1]. Tumor size is usually more than 4 cm, as it detects tumor late, unclear boundaries, and is characterized by rapid growth and destruction of surrounding structures and the destruction of the surrounding bony walls. Size of the tumor in this patient was also more than 4 cm.

The tumor develops rapidly, aggressively growing, destroying surrounding structures.

Our patient six months after surgery polyps in the nose and a normal finding in the nose and maxillary sinuses, comes with symptoms of pain in the jaw, allegedly after tooth extraction, which is similar to other authors described [10].

Osteolysis of the walls of the maxillary sinus, which is presented in the CT findings in this patient can point to the chronic inflammatory process, so one should be careful and think on SNUC [11].

Our patient was negative for EBV, as well as in most studies related to sinonasal undifferentiated carcinoma, although there are studies in Asia and Italy, where these patients were positive for the virus [3, 6].

The World Health Organization classifies this as a highly aggressive carcinoma tumor clinicopathologic distinctive, uncertain histogenesis, with locally aggressive growth, pleomorphic tumor cells with frequent necrosis, so it should be distinguished from other carcinomas as well as from olfaktor neuroblastoma [5].

Immunohistochemical antigens can vary from case to case. SNUC were immunoreactive with epithelial markers, including pan-cytokeratins (AE1/AE3), low molecular weight cytokeratins (CAM 5.2) and simple keratin (CKs 7, 8 and 19), or usually at a non CK4, CK5/CK6 and CK14 [1, 6, 12].

Sinonasal undifferentiated carcinoma is particularly difficult to treat [13-16]. Patients with resectable tumours are offered surgery followed by chemoradiation. Chemotherapy agents used include

carboplatin, cisplatin, and etoposide. Surgical resection typically involved extensive craniofacial resection with maxillectomy, orbital exenteration, and occasionally, neurosurgical involvement. Overall, outcomes of these patients were typical of those depicted in the literature: patients had high rates of recurrence, metastatic disease, and had very limited disease-free survival [17-18].

4. Conclusions

Sinonasal undifferentiated carcinoma is a rare and aggressive tumor of the maxillary sinus, requiring aggressive therapy. The exact cause of this cancer is not known, but there are cases of occurrence of cancer after the existence of polyps in the maxillary sinus and nose.

The optimal treatment for SNUC has yet to be determined.

It should be the early detection of tumors, timely surgical treatment in which is possible with radiotherapy and chemotherapy. Despite aggressive therapy, the prognosis for these patients is poor. Multimodal intensive therapy is recommended for all patients with SNUC.

The disease tends to recur in the same area from which it arises, and that treatment must focus on eliminating the disease via all available treatment modalities.

Based on our experience and a review of the recent literature, our future recommendations include: tumor registries for all patients with sinonasal malignancies, primary care physician education regarding warning signs and the importance of expedient diagnosis for all sinonasal malignancies.

We can continue to offer at least some hope to patients with this difficult disease.

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