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Nasosinusal Teratocarcinosarcoma: Case Report and Literature Review

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Abstract **Case Report**

Sinonasal cancers are the most common malignant tumors of the anterior skull base. Teratocarcinosarcomas are extremely unusual malignat tumors involving nasosinusal area, they are characterized by their poor prognosis and high likelihood of recurrence and potentiel intracranial extension. Because of its histological heterogeneity, the diagnosis is very difficult and therefore requires a sufficient sample to avoid erroneous diagnoses. The most commonly used treatment option is radical surgery resection followed by radiotherapy +/- chemotherapy. We report herein the case of 44-year-old woman, presented with sinonasal teratocarcinosarcoma revealed by nasal obstruction and intermittent epistaxis. The diagnosis was confirmed by a histopathological study of surgical specimen. Pre-treatment workup was negative for metastasis disease. She has received adjuvant radiotherapy and chemotherapy after radical endoscopic sinonasal surgery. A follow-up examination at 2 years showed no evidence of recurrence.

Keywords: Sinonasal teratocarcinosarcoma, Case report, woman, Surgery, adjuvant Radiotherapy+/- chemotherapy.

INTRODUCTION

Sinonasal teratocarcinosarcomas (SNTCSs) are a rare and unusual malignant neoplasm**,** that often arises paranasal sinuses. They are highly aggressive with high rate of recurrence and potential intracranial extension. Due to its heterogeneity and histological variety, the diagnosis of SNTCSs is very challenging, it is based on the presence of malignant epithelial elements and two or more malignant mesenchymal components (10). The adult is mainly affected by this lesion with significant male predominance [18]. The optimal treatment strategies are not well characterized because of the rarity of this tumor; in the literature, the most common treatment modality utilized is surgery and adjuvant radiotherapy +/- chemotherapy. We report the case of sinonasal teratocarcinosarcoma in 44 years old woman while discussing the epidemiology, diagnosis, and treatment of this rare entity.

CASE REPORT

A 44-year-old white woman, presented to the Bucco-maxillofacial Surgery Department in intercommunal hospital in Créteil with 1 month of nasal obstruction and intermittent epistaxis.

She has a 20-year history of smoking one pack a day. She reported three cases of cancer in her family; lung cancer in her father, testicular cancer in a sibling, and breast cancer in a maternal grandmother and two maternal aunts.

On examination, her vital signs were within normal range and the rest of the physical and neurological examinations were normal. A nasal speculum examination found a budding bleeding mass on contact. It occupied the entire right nasal cavity and caused a deviation of the nasal septum on the left. No palpable lymph nodes were observed.

A cervical computed tomography (CT) scan showed a homogeneous density opacity of the right ethmoidal sinus, measuring 3 cm in greater diameter and extending into the sphenoidal sinus.

Magnetic resonance imaging revealed expansive mass involving the right nasal cavity, ethmoidal and sphenoidal sinuses measuring 45 mm in maximal diameter with extension to the left ethmoid. There was no intracranial extension.

A biopsy with histological examination initially concluded the diagnosis of high-grade neuroblastoma.

Preoperative workup including positron emission tomography (PET) scan was negative for metastasis disease.

After systematic assessment, radical endoscopic sinus surgery was suggested to the patient. The tumor was removed completely without residual disease. The histopathological analysis of the surgical specimen confirmed the diagnosis of teratocarcinosarcoma.

Adjuvant treatment with concurrent chemoradiotherapy was validated by a multidisciplinary meeting and proposed to the patient.

We discovered on the pretreatment CT scan a pneumencephaly related probably to a bone breach. Unfortunately, the patient was admitted to the neurosurgical department and underwent further surgery that discovered a brain abscess.

One month after the surgery, the patient received a combination of concomitant radiochemotherapy with cisplatin at a dose of 54 Gy within a tumor bed delivered at 2 Gy per fraction in 5,4 weeks without treatment interruption. IMRT technique was used for treatment with acceptable G2 toxicity.

Three months after radiochemotherapy, clinical evaluation noticed the persistence of some side effects of radiotherapy such as mucositis and epiphora. The Head-and-neck magnetic resonance imaging (MRI) revealed an inflammatory lesion in the right orbital apex without residual disease. The following MRI showed the same radiological aspects.

No evidence of recurrence has been observed for over 2 years.

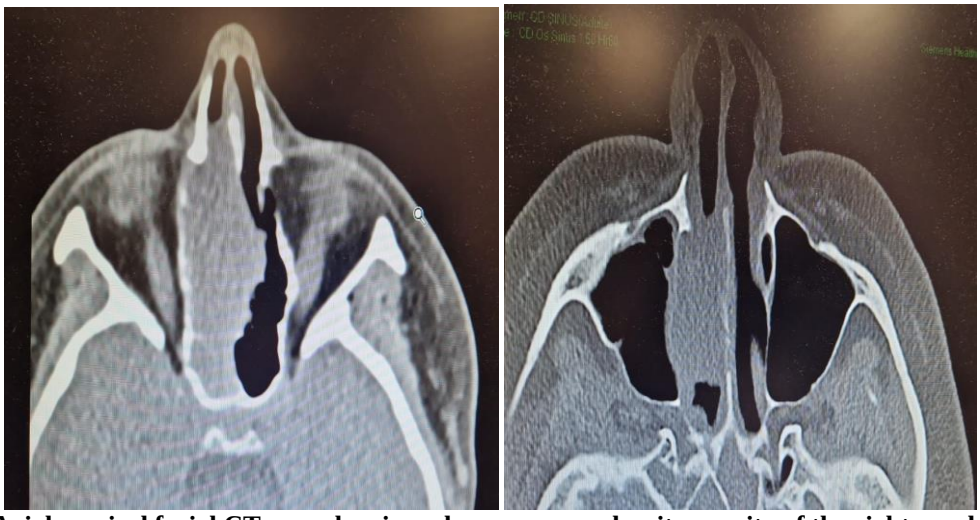

Figure 1: Axial cervical facial CT scan showing a homogeneous density opacity of the right nasal cavity and ethmoid sinus extending into the sphenoidal sinus

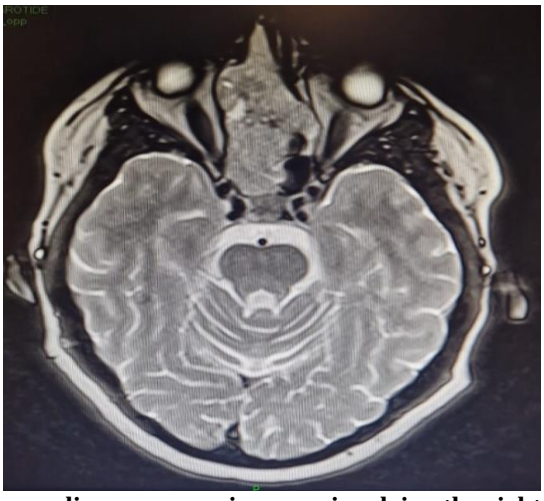

Figure 2: T2 weighted MRI revealing an expansive mass involving the right nasal cavity, ethmoidal and sphenoidal sinuses and extending to the left ethmoid

DISCUSSION

Epidemiology

Teratocarcinosarcomas are rare and highly aggressive malignant tumors [1]. It was previously designed as malignant teratomas and blastomas. In 1984, Heffner and Hyams referred these types of tumor to as teratocarcinosarcoma in view to describe the complex histological pattern [2, 3].

Sinosal cavity is the primary common site of teratocarcinosarcoma, although some may occur in other locations including the nasopharynx and oral cavity [4]. Intracranial extension is present in about 20.9% of cases [5].

The medical literature has reported less than 100 cases [1]. Most of them were adult with an average age of 35 years old and has an 8:1 predilection towards men [6] contrary to what is presented in our case report which the patient has 44 years old. Only five cases reported in children [8].

Diagnosis and Workup

Clinically, the most common symptoms are nasal obstruction and recurrent epistaxis. However, the patient can also present with anosmia, headache, visual changes, and neurological symptoms [7].

The course of the disease generally ranges from weeks to 2 years [6]. Conventional imaging is helpful but limited in the identification of histological type, comprehensive staging and post-therapeutic surveillance of TCS.

SNTCS usually lacks specific CT and MRI features; the images are similar to those of common malignant sinus tumors [6].

Histopathology

Histologically, SNTCSs are a combination of malignant teratoma and carcinosarcoma with a triphasic growth pattern including epithelial, mesenchymal, and primitive neuroectodermal components with teratoid elements [16].

The epithelial tissue is composed with both begnin and malignant squamous and gladular cells, they are usually immature or poorly differentiated and lack the apparent atypical organized features compared with typical squamous cell carcinoma and adenocarcinoma [17].

As for mesenchymal elements include fibroblasts, cartilage, bone, and smooth muscle; but one of the most components found in SNTCS sis the rhabdomyosarcoma.

Given the histological diversity of SNTCSs, the diagnosis is very challengning and therefore inadequate sampling as tissue biopsy alone may lead to erroneous diagnoses of olfactory neuroblastoma; as in our case; small cell carcinomas, immature teratomas, malignant transformations of teratomas, carcinosarcomas, or adenocarcinomas [14].

Treatment and Prognosis

The optimal approach to treatment remains unclear in this condition due to the lack of large-scale studies given the rarity of the disease.

Given its high rate of malignacy and low survival, SNTCSs requires an aggressive therapeutic approach; in almost of cases combining surgery with adjuvant radioterapy [4]. This association apperas to be the most commonly selected used in 62% of cases for which treatment data is available [4].

Nikita *et al*., have objectified by using pairwise log-rank test in their metaanalysis of 85 studies, that trimodality combining surgery with chemoradiation was also associated with significantly delayed time to death as compared to bimodality.

Moreover, in the previous studies, we have found that the highest survival rate (88.8%) and lowest recurrence rate (22.2%) were found in the group that underwent surgery with adjuvant focused radiation and chemotherapy [6].

The current paradigm of surgical treatment of SNTCSs is complete wide excision of the primary tumor with a margin of uninvolved tissue whenever possible. As for surgical treatment, according to the location and extension of tumor, the surgical approaches vary from craniofacial resection (18.6%), maxillectomy approach (12.8%) and rhinotomy (10.5%) [4].

Radiotherapy as an important therapeutic option has been described in management of SNTCSs; It is used systematically after surgery to eradicate residual disease. The average dose of radiotherapy used was 55 Gy, and the preferred technique adopted in the previous study was IMRT with goof effect. Two case reports by Tokunaga *et al*., and Peng *et al*., have utilized IMRT with good effect, with no recurrence at 2 and 3.5 years respectively [12].

As for chemotherapy, some studies suggested the importance of neoadjuvant chemotherapy prior to operation due to the anatomical complexity of the nasal sinus and the surgery alone cannot guarantee complete tumor resection [6]. While The role of adjuvant chemotherapy remains unclear and therefore its role is not well defined; There is some reports that adjuvant chemotherapy can lead to better survival outcomes (88.8% survival rate in addition to surgery and radiation vs 56,5% for surgery and radiation alone) [4].

The prognosis of SNTCSs is very poor with high risk of recurrence (its rate ranges from 25% to 55%) [6] and metastasis.

In a meta-analysis [13], 54 cases of SNTCS were analyzed and they found 67% of patients with initial surgical resection and 80% of patients primarily treated with radiotherapy had recurrence, metastasis, or were unresponsive to treatment. While 70% who had undergone combined surgery and adjuvant therapies, were survived more than 1 year and therefore an aggressive therapeutic approaches should be proposed as soon as possible in view to improve treatment outcomes [13].

CONCLUSION

Sinonasal teratocarcinosarcomas are uncommon and aggressive malignant skull base tumor with poor prognosis and frequent replace and metastasis. Atteinte féminine is unusual. Its management requires multidisciplinary combination of multiple therapeutic modalities ; radical surgery with adjuvant radiation +/- chemotherapy appears the adequate therapeutic approach but further research must be conducted to standardize management protocols for these tumors.

Conflict of interest: The authors declare no conflict of interest.

Informed Consent: The patient's informed consent to the publication of their data has been obtained.

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