Case Report

Sinonasal Teratocarcinosarcoma: A Case Report and Literature Review

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Abstract:
Sinonasal teratocarcinosarcoma is a rare type of malignancy which almost exclusively arises in the sinonasal tract, consisting of both the epithelial tissue and one or more connective tissue elements. This is a male predominant malignancy and most of the patient present with nasal obstruction and epistaxis but can also present with other symptoms depending on tumor extension into the adjacent structures like cranial cavity or orbit. Distant metastasis is rare but may involve spinal axis, cervical lymph nodes and respiratory tract. This paper includes a rare case report of Sinonasal teratocarcinosarcoma in a 70-year-old male, which was managed by surgical treatment and radiation therapy.

Key words: Sinonasal teratocarcinosarcoma**,** Distant metastasis, Surgical treatment, Radiation therapy

Introduction:

Sinonasal teratocarcinosarcoma is a rare type of malignancy which almost exclusively arise in the sinonasal tract, consisting of both the epithelial tissue and one or more connective tissue elements[1]. In the past, they were named as malignant teratoma, teratoid carcinoma, carcinosarcoma, blastoma, mixed mesodermal tumor etc.[2] In 1984, Heffner & Hyams first introduced the name teratocarcinosarcoma which explains the complex histological elements of both teratomas and carcinosarcomas[3,4]. This is a male predominant malignancy with male to female ratio of 7:1[5]. Most of the patient present with nasal obstruction and epistaxis but can also present with other symptoms depending on tumor extension into the adjacent structures like cranial cavity or orbit. Patient may present with headache, dizziness, focal neurological signs, and visual disturbances[6]. Distant metastasis is rare but may involve spinal axis, cervical lymph nodes and respiratory tract[7].

Case Report:

A 70-year-old male presented with epistaxis and nasal obstruction for 3 months and loss of vision of both eyes (Lt>Rt) for 2 months. Patient was hypertensive and on antihypertensive drugs. On examinations of nose, there was a mass present in the right nasal cavity. Ear and oral cavity examinations revealed normal findings. Clinically no cervical lymphadenopathy was found.

CT scan of Para nasal sinuses revealed a large right sided sinonasal soft tissue mass lesion with lobulated margin with destruction of lateral bony wall of right bony orbit, right sided greater wing of sphenoidal bone, bony wall of nasal septum. The lesion occupied right and left side of nasal cavity, right maxillary antrum, both sided ethmoidal sinuses, right retrobulbar space of orbit, floor of right frontal bone and medial wall of right bony orbit. Right sided nasal cavity was almost obliterated and nasal septum was deviated to left side. There was local extension into the inferior surface of right frontal lobe of brain. Bony outlines of left sided maxillary and frontal sinuses were normal. No extension into the nasopharyngeal region was noted.

Patient underwent endonasal endoscopic anterior craniofacial excision of mass under general anesthesia.

Histological Examinations:

Microscopy: Sections show stratified squamous epithelium with underlying tissues having a tumor

composed of small polygonal cells having round hyperchromatic mildly pleomorphic nuclei and very scantly cytoplasm. The cells are arranged in nests and trabecular pattern.
Occasional glandular structures are also seen. Focal areas show rosette formation and necrosis (++), tumor emboli (+), mitotic rate 20/10 HPF.

Immunohistochemical study:
Pan cytokeratin- Positive in most of the cells.
Synaptophysin- Positive in many tumor cells and negative in many cells.
Ki67- Positive in all the cells (MIB index-100%)
GFAP- Negative
Patient was treated with post-operative radiotherapy by IMRT technique. Total 6000 cGy radiation is given in 30 fractions. 7 fields were used. On treatment radiation induced toxicities were manageable. Now patient is scheduled for two monthly follow ups.

Discussion:
Sinonasal teratocarcinosarcoma is a rare variety of malignancy of sinonasal tract that is composed of high grade carcinosarcoma and teratoma[4]. This type of malignancy comprises both benign and malignant component[8]. In the past there were so many nomenclatures, but in 1984 Heffner and Hyam proposed a unique terminology called "Teratocarcinosarcoma" through their review of 20 cases[2].
This is a male predominant malignancy with male to female ratio is 7:1[5]. Usually, it occurs between the age group of 18 to 79 years with median age group of 60 years[3]. Most commonly sinonasal teratocarcinosarcoma arises from nasal cavity, maxillary, and ethmoidal sinuses, but a single case of sphenoidal sinus teratocarcinosarcoma has also been reported[9]. Sinonasal teratocarcinosarcoma patients most commonly present with nasal obstruction and epistaxis. Other presentations may include headache, dizziness, proptosis, visual problem, and neurological symptoms due to local extension of disease[6].
Histopathologically sinonasal teratocarcinosarcomas are characterized by various combinations of benign and malignant elements. It includes bone, muscle, and cartilage along with glandular, neuroendocrine, neuroepithelial and epidermoid tissues[10]. Histopathology is the cornerstone of diagnosis. Detailed and thorough examination of histological specimen is essential for diagnosis[11] along with appropriate imaging like CT scan or MRI. Carcinomas, sarcomas, neuroendocrine tumors, lymphoma, and melanoma should be included in histological differential diagnosis[5]. Immunohistochemistry also helps to confirm the diagnosis and exclude differential diagnosis.
Multidisciplinary approach in the form of surgical resection, radiation therapy and chemotherapy are needed to treat such malignancy[12]. Use of chemotherapy is very few and has been used in some extensive and recurrent cases. Due to the rare nature of the tumor, it is difficult to collect reasonable evidence of an improved cure rate following chemotherapy. It is proved that patients who did not receive postoperative adjuvant radiotherapy had a higher rate of recurrence when compared to those who had radiotherapy[13,14]. Distant metastases are rare as the tumor is locally aggressive, but lung metastases have been reported[15]. In metastatic setting, palliative chemotherapy may be given to improve survival as metastatic tissue often contains sarcoma components. Three- and five-years survival rate is about 30% and 20% respectively[3].

Conclusions:
In conclusion, Sinonasal teratocarcinosarcomas are extremely rare variety of malignancy and recurrence rate is also high. So, according to the consensus of recently published article, we adopted excision of the mass followed by adjuvant radiotherapy by IMRT technique. In such cases, regular follow up is mandatory to detect early recurrences.

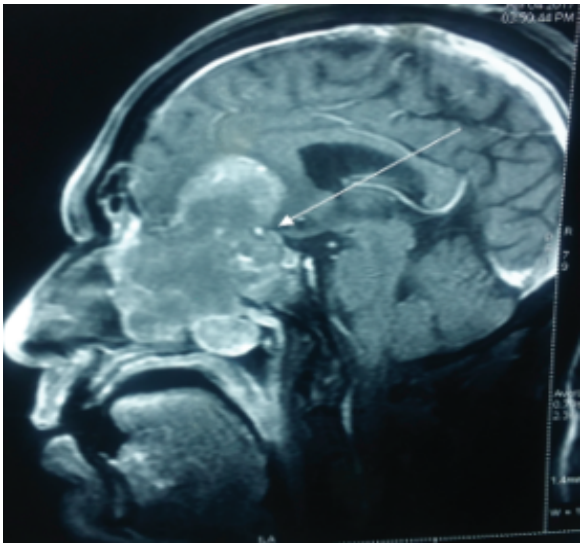


Figure-1: Pre-operative image



Figure-2: Pre-operative image

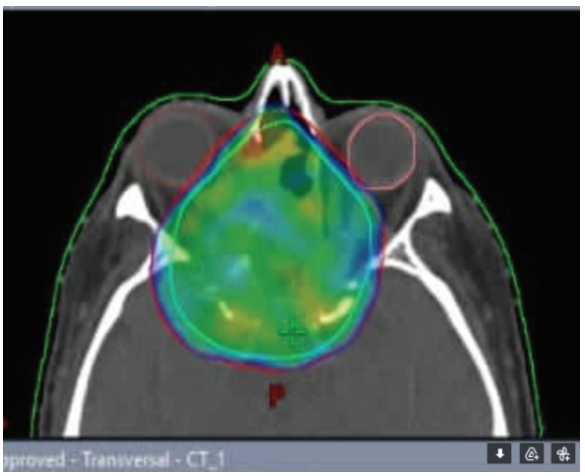


Figure-3: Radiotherapy planning coverage

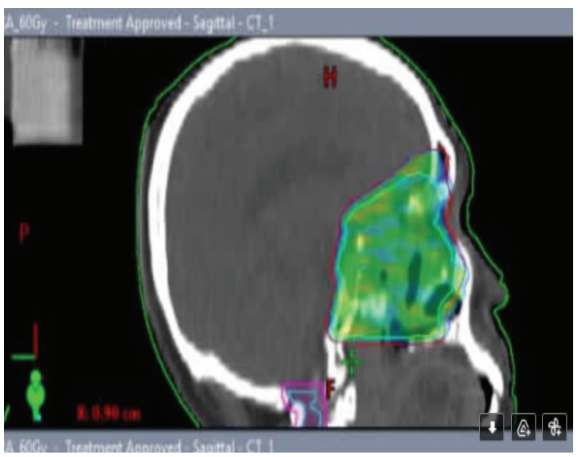


Figure-4: Radiotherapy planning coverage

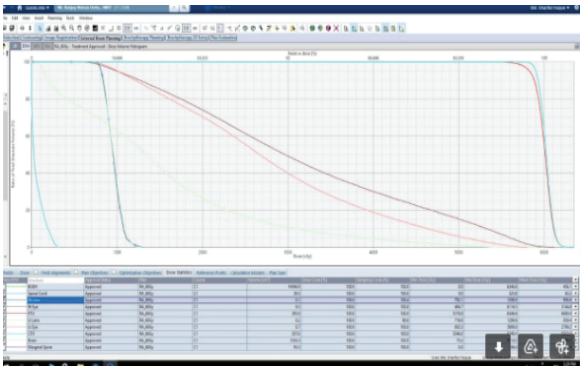


Figure-5: Dose volume histogram (DVH)

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