CASE REPORT

Sinonasal teratocarcinosarcoma of the ethmoid and paranasal sinus: A rare neoplasm

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Sinonasal teratocarcinosarcoma (SNTCS) is a very rare and aggressive malignant neoplasm histologically characterized by the combination of one or many components of epithelial and mesenchymal elements. Here, we report a SNTCS in a 60-year-old man involving posterior nasal and nasopharyngeal wall extending into left ethmoidal sinus. The patient complained of bleeding from nose, nasal obstruction, and generalized weakness for last two months. Tumor was completely removed by Caldwell-Luc operation and postoperative radiation therapy was given. The follow-up of the patient for two years has shown no evidence of recurrence or metastasis.

Key words: Clinical presentation, histopathology, sinonasal teratocarcinosarcoma

INTRODUCTION

Sinonasal teratocarcinosarcoma (SNTCS) is a distinctly rare tumor characterized by variegated histologic features and has combination of both epithelial and mesenchymal components.[1,2] Malignant tumors which consisted of carcinoma and teratoma were previously reported as teratoid carcinoma, malignant teratoma, blastomatous tumors, and blastoma. It was first described by Shanmugaratnam *et al.* in 1983 and was aptly termed as "Teratocarcinosarcoma" by Heffner and Hyams in 1984.[3,4] Probably, this is the first case report of SNTCS from eastern India. By reported accounts, SNTCS is a highly malignant tumor displaying rapid aggressive growth. Prognosis is poor. Five-year survival rate is 30%–50% with 60% mortality beyond three years.

CASE REPORT

A 60-year-old man presented with occasional bleeding from nose and nasal obstruction for last two months. He also had generalized weakness and episodic headache. His hemoglobin level was 11.7 gm/dl at the time of presentation. Other routine hematological and biochemical investigations were normal. X ray of paranasal sinus showed a mass in the posterior nasal cavity. A computed tomography (CT) scan revealed a solid tumor with isodensity. The mass was in the posterior nasal cavity and nasopharyngeal wall extending into left ethmoidal sinus [Figure 1]. Diagnostic imaging suspected of sinonasal malignancies such as poorly differentiated carcinoma, adenocarcinoma, and malignant lymphoma.

The patient was operated with all antiseptic measures under general anesthesia during Caldwell-Luc operation. Cheek was retracted and an incision was made above the canine teeth in the buccogingival sulcus. It was about 3–5 cm long and was placed high enough to prevent damage to the dental roots. A part of the anterior wall of left antrum was removed with a chisel and hammer to make an opening allowing an access into the antral cavity. The mass was removed with Luc's forceps. An intranasal antrostomy was done. The wound was sutured with absorbable catgut suture. The specimen was sent for biopsy. Multiple gray–white tissue bits, largest measuring 3.5 × 3 × 2.5 cm received grossly for histopathological examination. Microscopically, the tumor mass showed variegated histologic architecture. Benign and malignant components of epithelial and mesenchymal tissues were detected. The epithelial portion of the tumor was composed of benign and malignant squamous epithelium, glandular structure, and primitive neuroectodermal/blastemal cells [Figures 2 and 3]. Clear cell squamous epithelia were present in the epithelial component of the neoplasm. Teratoid components included this "fetal appearing" clear cell squamous epithelium and primitive neuroectodermal/blastemal cells. In the mesenchymal components; spindle cell sarcoma was noted [Figure 4]. Many mitoses including atypical cells were detected. MIB1

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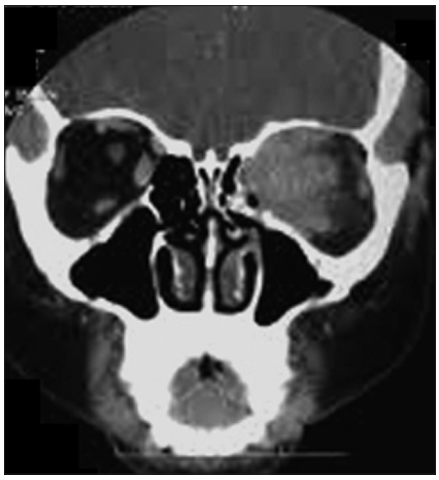

Figure 1: CT scan showing a mass involving the left paranasal sinus and ethmoid sinus

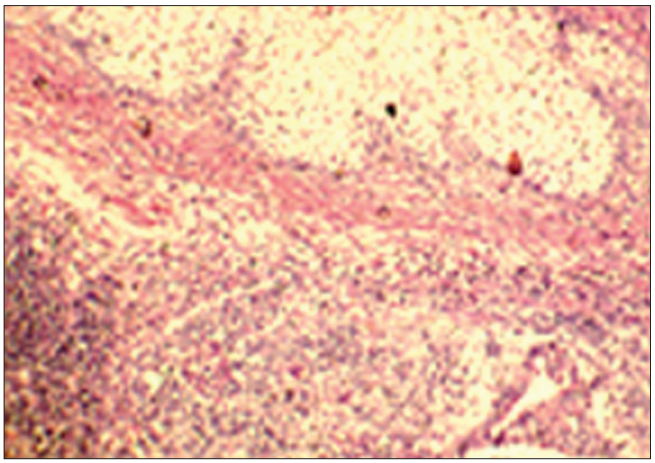

Figure 2: Photomicrograph showing fetal appearing (clear) squamous epithelial cells, glandular components, and primitive neuroectodermal/blastemal cells in the background. [H and E, × 100]

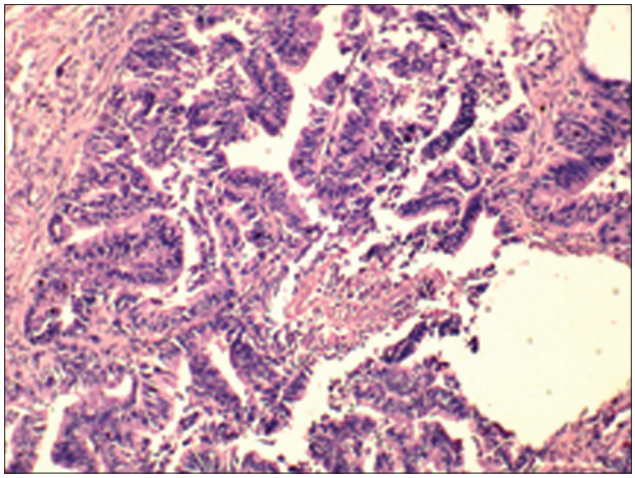

Figure 3: Photomicrograph showing malignant epithelial cells arranged in glandular pattern. [H and E, × 400]

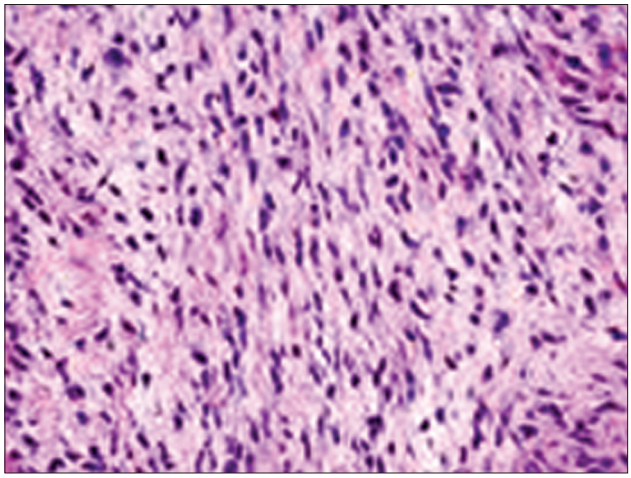

Figure 4: Photomicrograph showing spindle cell sarcoma. The tumor cells have nuclear aypia and mitosis. [H and E, × 400]

(Ki 67) positivity was detected in 25% of the mesenchymal components and 37% of the epithelial components of the malignant tumor, whereas the clear cell epithelia revealed MIB1 (Ki67) positivity in 5% of cells. Patient was treated with radiotherapy (60 Gy) followed by surgery. A whole body CT scan was done at six months interval to detect any recurrence or metastasis. Follow-up examination for two years after combined surgery and radiotherapy has shown no evidence of recurrence or metastasis.

DISCUSSION

Malignant tumors having teratoma and carcinoma were reported previously as teratoid carcinoma, malignant teratoma, or blastematous tumors.[5] Heffner and Hyams in 1984 first suggested the tumor to be called as teratocarcinosarcoma in order to describe the complex histological pattern of these neoplasms. Histologically, it is different from true carcinosarcoma which consists of a single malignant epithelial and a single malignant mesenchymal component, whereas SNTCS has one or many epithelial and mesenchymal components (both benign and malignant).[6] Variegated architecture and tissue heterogeneity are characteristics of this malignancy. The malignant epithelial component includes squamous cell carcinoma and adenocarcinoma. "Fetal-type" clear cells, squamous epithelium, and immature neuroepithelium represent important histologic characteristics useful in diagnosis.[3,7] The present case had characteristic features of SNTCS including epithelial and mesenchymal elements. Though immature neuroepithelium (primitive neuroectodermal/blastemal) was present, differentiated neuroepithelial cells with rosette formation were not evident. Despite several studies, the histogenesis of this tumor remains controversial. Heffner and Hyams postulated that the tumor originates from olfactory membrane due to presence of neural tissue. Some authors believe that SNTCS probably originates from primitive embryonic tissue or immature pleuripotential cells.[8] A histogenetic origin from a multipotential adult

somatic stem cell with divergent differentiaton has been favored over a germ cell origin. This assumption has been based on the lack of germ cell elements and, until recently, the absence of demonstrable amplification of 12p in tumor cells.[9] Ultrastructurally, the primitive cells had many neural processes with parallel microtubules. Tumor cells showing squamous cell differentiation were characterized by desmosome-like junction and intracellular tonofilaments. Some of the stromal spindle cells had actin filaments with dense patches and dense core granules.[10]

Shubhada *et al*., observed chemotherapy-induced neuronal maturation in sinonasal teratocarcinosarcoma. In that case, the tumor was excised after four cycles of neo-adjuvant chemotherapy. On microscopic examination, it showed similar epithelial and mesenchymal components as the pretreartment biopsies. However, the primitive neuroectodermal component displayed extensive neuronal maturation. The undifferentiated neuroectodermal cells were completely absent in the postchemotherapy specimen. This case highlighted morphologic evidence of chemotherapy-induced maturation in the neuroectodermal component within SNTCS.[11]

In a study by Budrukkar *et al*., disease recurred in 11 out of 14 patients, with a median time to recurrence of seven months. Multimodality treatment, in the form of a combination of surgery, radiation therapy, and chemotherapy, appears to be the optimal approach.[12] A combination of radiotherapy and surgical treatment offers the best five-year survival rate (50%); followed by only surgical treatment (47%). In the recurrent or metastasis lesion, adjuvant chemotherapy may improve the survival rate since the metastatic tissue often contains sarcomata components.

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