INTRODUCTION
Teratocarcinosarcoma is an extremely rare, highly aggressive, and rapidly growing malignant neoplasm. The tumour is variously termed as malignant teratoma, blastoma, teratocarcinoma or teratocarcinosarcoma. The tumour shows both epithelial (skin and lining of internal organs) and mesenchymal (bone, cartilage and fat) elements of the tissues. The tumour components include fetal clear cell squamous epithelium derived from ectoderm. Glandular tubular structures and ciliated columnar epithelium are derived from endoderm. Fibroblasts, striated muscle, smooth muscle, cartilage and osteoid matrix are derived from mesoderm. The carcinoma component exhibited mostly adenocarcinoma and squamous cell carcinoma, whereas the sarcoma component mostly exhibited rhabdomyosarcoma, leiomyosarcoma, and fibrosarcoma.

So far about 65 cases have been reported in the literature. Patients are exclusively adults, with a mean age of 60 years and approximate 8:1 male predominance. It often tends to occur in the nose, pharynx and sinus areas, but tumour in other parts of the body has also been recorded. Clinical features are determined by the size and location of the tumour. The most common complaints at presentation include nasal obstruction, epistaxis, facial pain, headache, proptosis and visual field deficits. The lesion is often misdiagnosed due to infrequency and the complex phenotypic composition. This tumour can be diagnosed by histopathology accompanied by immunohisto-chemical study for cytokeratin, epithelial membrane antigen, vimentin, smooth muscle actin, S-100 protein, neuron-specific enolase, synaptophysin, chromogranin, desmin, myosin and myoglobin. Fluorescent in situ hybridization evaluation can be used for diagnosis. Microscopically, the tumour shows epithelial nests, net of angioma-like fibrous connective tissues, small round and spindle cells, glandular, squamous-like cells, and cells of rhabdomyoblastic differentiation.

The high rate of local recurrence and distant metastasis is indicative of its highly aggressive biologic behaviour. Due to its aggressiveness, 3 years survival is approximately 50% with aggressive therapy. The diagnosis of teratocarcinosarcoma is based on the demonstration of teratoid and carcinosarcomatous features. Conventional cross-sectional imaging is helpful in the comprehensive staging and post-therapeutic surveillance of teratocarcinoma.

To the best of our knowledge, it is the first case of this disease found in this part of the world.

CASE REPORT
A 20 years old Afghan male was suffering from left sided nasal obstruction with headache for one year. On examination, patient had reddish, fleshy mass in left nasal cavity. Carotid angiography was inconclusive so incision biopsy was taken from nasal mass and it was reported as teratocarcinosarcoma. CT scan of PNS was done which showed heterogeneous mass in nasal cavity, paranasal sinuses with intracranial extension. The patient was operated. The approach was a combination of lateral rhinotomy with trans-basal sub-frontal craniotomy. He was treated by Co 60 and received a tumour dose of 56 GY radiation. He is being followed regularly every 2 months after his final radiotherapy session and he is disease-free so far.

Key words: Sinonasal teratocarcinosarcoma. Teratoid carcinosarcoma. Nasal polyp.
healthy physique. On examination, patient had a reddish, fleshy mass in left nasal cavity. It was sensitive to touch. The mass was also visible in nasopharynx. There was mild proptosis on the left side. There was retracted tympanic membrane on the left side. Rinne test was negative on left ear while other ear was normal. His vision was normal. His rest of ENT, cranial nerves and systemic examination was unremarkable. Carotid angiography was inconclusive so incision biopsy was taken from nasal mass and it was reported as Teratocarcinosarcoma. Then patient was further investigated and beside routine investigation ultrasonography of abdomen especially of testicles was normal. CT scan paranasal sinuses showed heterogeneous mass in nasal cavity, paranasal sinuses with intracranial extension (Figure 1). This was discussed with Neurosurgeons and on their advice MRI was done which also showed the same extension of lesion as in CT scan. As the lesion was extending into middle cranial fossa, a combination of lateral rhinotomy with trans-basal sub frontal craniotomy approach was adopted (Figure 2). The patient was operated in 6 hours time and disease was cleared from intracranial part by Neurosurgeon using microscope and from extracranial part by ENT surgeons. The specimen was again sent for histopathology study (Figure 3). Then the patient was referred to Oncologist for chemoradiotherapy. He was treated with Co 60 and received a tumour dose of 56 Gy radiation. The patient was rechecked one month after completion of radiation. He had no special symptoms except nasal dryness. He is being followed regularly every 2 months after his final radiotherapy session and he is disease-free so far.

DISCUSSION

Sinonasal-teratocarcinosarcoma (SNTCS) is a high-grade polymorphous tumour. Its unusual histological features make management difficult.4 The differential diagnosis of SNTCS include poorly differentiated carcinomas, sinonasal undifferentiated carcinoma, adenocarcinoma, neuroendocrine carcinoma, rhabdomyosarcoma, synovial sarcoma, olfactory neuroblastoma and malignant mixed salivary gland tumours.2,5 The cornerstones of treatment for sinonasal teratocarcinosarcoma include surgical resection or radiation therapy with 45-70 Gy or a combination of both. Chemotherapy is rarely effective in this condition. Despite intensive treatment, the prognosis is still poor.2-4 In some of the cases, the tumour was totally resected via the craniofacial approach and the patient was given postoperative chemotherapy. Extensive tumour necrosis, rapid growth and local destruction are the prominent features of this tumour. Similarly, in another case, Denker-Watsuji operation was performed, and the patient was treated with a combination of radiation therapy and chemotherapy.1,4 Distant metastasis of SNTCS is seldom described because of the invasive character of the cancer.3,6 Metastasis to the lungs and airways can cause obstruction of the main bronchi creating challenging dyspnea.7
The most common cause of treatment failure is local recurrence. The high rate of local recurrence and metastasis is indicative of its highly aggressive biologic behaviour. An aggressive elective neck dissection should be performed in the early disease stage, and more attention should be given to the soft tissue surrounding any possible lymphadenopathy. This may decrease the risk of lower cervical lymph node or distant metastasis in patients with sinonasal-teratocarcinosarcoma. Almost half of the patients die of tumour within 3 years of diagnosis, despite aggressive therapy. Seventy percent of the patients who survived for more than one year had the initial therapeutic regimen of combined surgery and adjuvant therapies, suggesting that aggressive therapeutic approaches may improve the treatment outcome. Adjuvant chemotherapy with Cisplatin, Etoposid and Ifosfamid can be given with regard to the major components of this heterogeneous tumour.

REFERENCES


