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Pathology

SINONASAL TERATOCARCINOSARCOMA – A RARE CASE REPORT

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ABSTRACT Sinonasal Teratocarcinosarcoma (SNTCS) is a very unusual and aggressive neoplasm characterized by a histologic combination of malignant teratoma and carcinosarcoma with a triphasic growth pattern including epithelial, mesenchymal and primitive neuroectodermal components. This neoplasm is mostly seen in adults with a mean age of 55 years. Here, we report a SNTCS in a 17 year old male presented with left sided nasal mass for last 6 months which was surgically resected. Histopathology and immunohistochemistry confirmed the diagnosis of sinonasal teratocarcinosarcoma. SNTCS is highly malignant and locally aggressive. About 60% patients do not survive beyond 3 years. Total excision and extensive sampling are necessary to reach the diagnosis. Early diagnosis and management can give a better prognosis.

KEYWORDS : malignant teratoma, carcinosarcoma, teratocarcinosarcoma

INTRODUCTION

Sinonasal teratocarcinosarcoma is a rare complex malignant neoplasm of sinonasal tract and consists of various carcinomatous and sarcomatous elements, including immature epithelial, neuroepithelial, and mesenchymal tissues resembling immature teratoma. SNTCS occurs mainly in the nasal cavity and paranasal sinuses, although tumours occurring in other locations including the nasopharynx and oral cavity have been described[1,2,3]. Previously described as teratoid carcinosarcoma, malignant teratoma or blastoma. It was first described by Shanmugaratnam et al. in 1983 and was aptly termed as "Teratocarcinosarcoma" by Heffner and Hyams in 1984[4,5]. SNTCS constitutes less than 1% of all cancers and approximately 3% of all malignancies of Head and Neck region[6]. Till date, there are <100 cases reported in english literature. It is almost exclusively seen in adults with only 3 pediatric cases being reported[7]. By reported accounts, SNTCS is a highly malignant tumour displaying rapid aggressive growth. Prognosis is poor. Five year survival rate is 30-50% with 60% moratlity beyond three years.

CASE SUMMARY

A 17 year old male patient presented with a left sided nasal mass with a non healing ulcer in the ala of nose for 6 months associated with nasal intonation of voice and loss of sensation of smell.

CECT reveals large soft tissue density lesion in left maxillary sinus causing bony erosions and extending to the nasal cavity, choana, nasopharynx, oropharynx and to the infratemporal fossa -- suggestive of SINONASAL MALIGNANT MASS[fig.1].

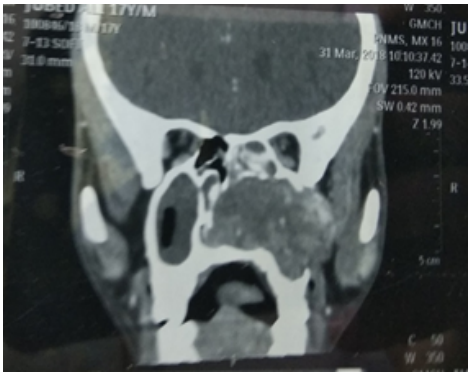

Fig – 1 CECT: mass in left maxillary sinus

Left lateral rhinotomy with medial maxillectomy with excision of nasal mass was done and the specimen was sent for histopathological examination.

HISTOLOGIC FEATURES

GROSS: Received a lobulated mass measuring (9 x 7 x 5.5) cm3 [fig.2a]. Cut sections are whitish solid and homogenous with few haemorrhagic spots at the periphery[fig.2b].

Fig-2a: gross morphology of specimen

fig-2b: cut section of specimen

MICROSCOPIC EXAMINATION

Multiple sections submitted show intimate admixture of gland lined by atypical columnar epithelium and nests of squamous epithelium at places surrounded by teratomatous elements in the form of cartilage, immature neural tissue, mature glial tissue and sheets of undifferentiated malignant round cells.

On higher magnification, focally glands show features of well differentiated adenocarcinoma and nests of immature squamous epithelium also seen. Brisk mitotic activity noted in the undifferentiated areas.

Immunohistochemistry shows strong neuron specific enolase and CD99 positivity and focal positivity for synaptophysin and chromogranin in small round cells.

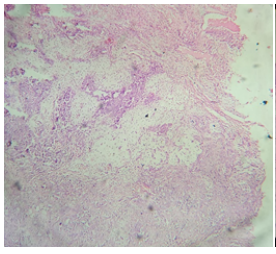

fig-3 100x chondroid differentiation

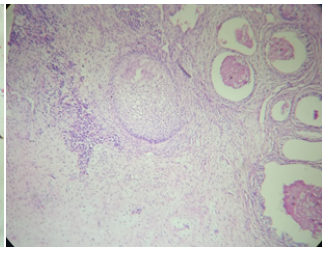

fig-4 100x nest of immature squamous cells

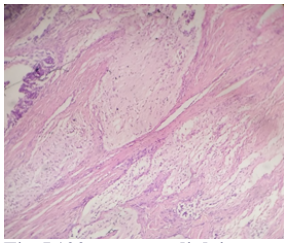

Fig-5 100x mature glial tissue

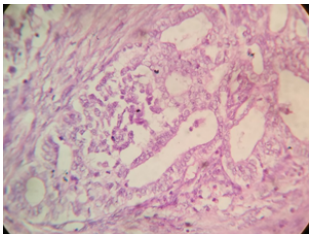

fig-6 400x adenocarcinoma

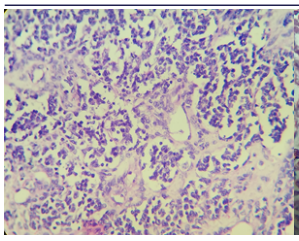

Fig 7 400x undifferentiated small round cells

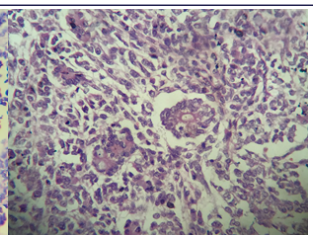

fig 8 400x rossettes

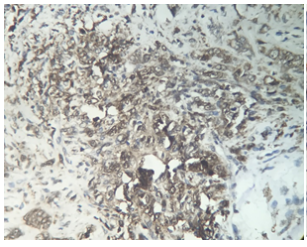

Fig-9 NSE

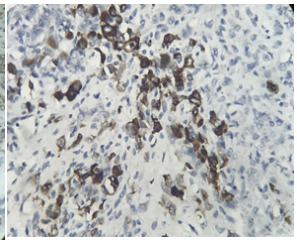

Fig-10 Synaptophysin

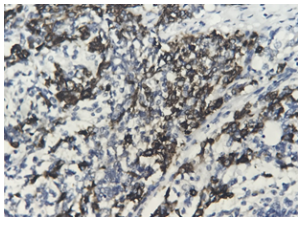

Fig 11 chromogranin

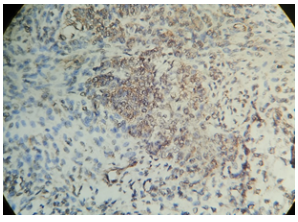

Fig 12 CD99

DISCUSSION

Sinonasal teratocarcinosarcoma is a malignant sinonasal neoplasm with combined histological features of teratoma and carcinosarcoma, lacking malignant germ cell component. Benign and malignant epithelial, mesenchymal and neural elements are typically present, including immature tissue with blastomatous features.

The term *sinonasal teratocarcinosarcoma* was first released in the World Health Organization 2005 version of *Pathology and Genetics of Head and Neck Tumors*[6]. SNTCS mainly located in the nasal cavity, followed by the ethmoid sinus and the maxillary sinus[2]. lntracranial extension occurs in approximately 20% of cases. This neoplasm mostly seen in adults, with a reported age range from 18 to 79 years with a mean age of 55 years, and a strong male predominance with a 7:1 male to female ratio[8].

Most common presenting symptoms are nasal obstruction and epistaxis. Imaging studies show a nasal cavity mass with opacification of paranasal sinuses and frequent bone destruction.

Heffner and Hyams postulated that the tumour 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[9].

There are limited reports in the literature on the cytogenetic abnormalities. These abnormalities include extra copies of chromosome 12p in a subpopulation of neoplastic cells in a hybrid case that also exhibited foci of yolk sac elements[10].

Tumour tissue is firm to friable, with a variegated reddish purple to brown appearance. When present, the surface mucosa is often ulcerated, and areas of necrosis and haemorrhage are evident at the cut surface.

Teratocarcinosarcoma is composed of an admixture of epithelial, mesenchymal and neuroepithelial elements. The epithelial components include keratinizing and non-keratinizing squamous epithelium, pseudostratified columnar ciliated epithelium and glandular/ductal structures. An important diagnostic feature is the presence of nests of immature squamous epithelium with clear so-called fetal-appearing cells[5]. The most represented mesenchymal elements are spindle cells with features of fibroblasts or myofibroblasts, but areas of rhabdomyoblastic, cartilaginous, osteoblastic, smooth muscle or adipocytic differentiation can be seen, with appearances ranging from benign to frankly malignant. The neuroepithelial component consists of a proliferation of immature round to oval cells either in solid nests or within a neurofibrillary background, sometimes with rosette formation.

Inadequate sampling may lead to erroneous diagnosis of olfactory neuroblastoma, squamous cell carcinoma, undifferentiated carcinoma, adenocarcinoma, malignant salivary gland type tumours and adenosquamous carcinoma[11].

The average survival is <2 years with 60% of the patients not surviving beyond 3 years. Recurrence is common, usually within 3 years.

CONCLUSION

Teratocarcinosarcoma is a aggressive sinonasal tumour, with frequent lymph node and distant metastasis. Total excision of the tumour and aggressive sampling for histopathological examination is necessary to avoid erroneous diagnosis.

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