Original Research Paper

Volume - 11 | Issue - 12 | December - 2021 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Histopathology

SINONASAL TERATOCARCINOSARCOMA: A RARE ENTITY

Varsha Dhume — Associate Professor, Department of Pathology, Topiwala National Medical College, Mumbai.

Kavita Pawar* — Assistant Professor, Department of Pathology, Topiwala National Medical College, Mumbai. *Corresponding Author

Aniket Meshram — Assistant Professor, Department of Pathology, Government Medical College, Nagpur.

ABSTRACT Sinonasal teratocarcinosarcoma is a rare nasal cavity tumor of uncertain histogenesis and having combined features of teratoma and carcinosarcoma. Here, we are presenting two cases of sinonasal teratocarcinosarcoma occurring in young males (22 and 35 years). Both presented with nasal cavity masses; one extending up to soft palate & hypopharynx and 2nd involving maxillary sinuses. Grossly, one of the masses has whitish firm, congested whereas other one was reddish friable hemorrhagic. Both were surgically resected. Paraffin-embedded sections were made and stained with hematoxylin and eosin. Microscopic examination revealed variegated appearance. Epithelial component comprised of malignant glandular epithelium as well as immature squamous epithelium. Whereas sarcomatous component revealed spindle shaped cells with high N/c ratio and hyperchromatic nuclei. Focal areas showed immature neuroepithelium. These findings clearly suggest the diagnosis of sinonasal teratocarcinosarcoma.

Sinonasal teratocarcinosarcoma is highly malignant and locally aggressive. About 60% of the 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, sinonasal mass, teratocarcinoma

INTRODUCTION

Sinonasal teratocarcinosarcoma (SCTCS) is an extremely rare and locally aggressive lesion arising from the nasal cavity and paranasal sinus in adults. First described by Shanmugaratnam et al as malignant teratoma in 1983 and then as teratocarcinosarcoma by Heffner and Hyams in 1984[1,2]. Till date less than 150 cases published in the literature[3].TCS is a complex malignant sinonasal neoplasm combining features of teratoma and carcinosarcoma. Patients are exclusively adults, with age ranging from 18-79 years (mean 60 years). There is a marked male predominance[2]. It almost exclusively arises in the ethmoid sinus and maxillary antrum. Patients present with a short history of nasal obstruction and epistaxis. Bone destruction may be seen. Tumours are usually bulky, soft to rubbery, and red-tan to purple. There are multiple tissue types derived from two or three germ layers, exhibiting variable degrees of maturity[1]. Tumor is composed of epithelial, sarcomatous and primitive components. The epithelial component is usually made up of keratinizing and nonkeratinizing squamous epithelium, pseudostratified columnar ciliated epithelium, and glandular structures lined by either cuboidal or columnar cells that may show mucous differentiation. Nests of immature squamous cells containing clear cells (fetal-appearing) are a common finding and an important. Teratocarcinosarcomas are highly malignant. The average survival is less than 2 years, with 60% of the patients not surviving beyond 3 years. Recurrences usually appear within 3 years. Here we are presenting 2 cases of SCTCS in a 35 year and 22-year-old male and discuss the clinical and histopathological features of this tumor.

CASE REPORT

Two male patients, 35 and 22 years old respectively presented with left sided nasal obstruction and recurrent episodes of epistaxis. In both the cases, CT scan of paranasal sinuses showed non-enhancing polypoidal masses in nasal cavity one; extending up to soft palate & hypopharynx and 2nd involving maxillary sinuses. The mass was surgically resected and fixed in 10% formalin. Grossly, one of the masses has whitish firm, congested whereas other one was reddish friable hemorrhagic. Paraffin-embedded sections were made and stained with hematoxylin and eosin. Microscopic examination revealed variegated appearance. Epithelial component comprised of malignant glandular epithelium as well as immature squamous epithelium. Whereas sarcomatous component revealed stromal cells which were spindle shaped with high nuclear: cytoplasmic ratio and hyperchromatic nuclei. Focal areas showed immature neuroepithelium. These findings clearly suggest the diagnosis of sinonasal teratocarcinosarcoma.

DISCUSSION

It is a rare, aggressive and highly malignant sinonasal neoplasm combining features of teratoma and carcinosarcoma. There are < 150 cases reported in literature till now[3]. This tumor mostly occurs in sinonasal tract. However, Fatima et al have been reported few cases in nasopharynx, orbit, posterior pharyngeal wall and tongue in her study[5]. Patients are exclusively adults, with age ranging from 18-79 years. SNTCS also seen in neonate by B. Rosenberg et al[6]. There is a marked male predominance.

SNTCS patients usually present with nasal obstruction and recurrent epistaxis. Nasal cavity on examination shows reddish brown, friable masses with necrotic areas as seen one of our cases[7].

Histological examination, reveals multiple tissue types derived from two or three germ layers, exhibiting variable degrees of maturity. The epithelial component is usually made up of keratinizing and nonkeratinizing squamous epithelium, pseudostratified columnar ciliated epithelium, and glandular structures lined by either cuboidal or columnar cells that may show mucous differentiation. Nests of immature squamous cells containing clear cells (fetal-appearing) are a common finding and an important diagnostic clue by some authors[2]. Pai and Naresh reported fetal squamous cells in two cases out of their four cases whereas we got in both the cases[8]. Though, it may represent a very small compound of the entire neoplasm. It is quite helpful in diagnosis when found. The carcinomatous component is usually glandular, but sometimes squamous. Neuroepithelial elements with rosettes and neuroblastoma-like areas are present in most instances[9]. The mesenchymal areas range from immature tissues (such as cartilage) to sarcomas (such as rhabdomyosarcoma and fibrosarcoma. SNTCS has diverse histology So, to achieve the correct diagnosis adequate sampling of tumor tissue is needed. One of cases, presented as hematoma and thorough sampling of resected tissue helped us to arrive to a correct diagnosis.

The important differential diagnosis which should be considered is carcinosarcoma and it differs from SNTCS as it consists of both malignant epithelial and mesenchymal components whereas SNTCS has many such components which may be benign or malignant[10]. Other differential includes olfactory neuroblastoma, sarcomatoid carcinoma, squamous cell carcinoma, undifferentiated carcinoma, adenocarcinoma, malignant salivary gland type tumors and adenosqu amous carcinoma and other sarcomas which can be ruled out by proper sampling of the tumor tissue[8].

The IHC can shows variable picture depending on the histological components. The undifferentiated/primitive component often shows positive immunoreaction for CD99 and occasionally synaptophysin and S-100 protein[8].The spindle cell component is consistently positive for vimentin, and sometimes desmin, myoglobin, and glial fibrillary acidic protein. The neuroepithelial component is positive for neuron-specific enolase and occasionally chromogranin, alfa-fetoprotein, and

cytokeratin. The epithelial component is positive for cytokeratins, epithelial membrane antigen, and occasionally S-100 protein and glial fibrillary acidic protein. Immunohistochemical profile can be variable and there is no single diagnostic marker.

These tumors are locally aggressive, rapidly invade soft tissue and bone as well as orbit and cranial cavities. They have also potential to metastasize to regional lymph node and distant sites, mainly lungs[4]. Treatment for SNTCS is radical excision followed by radiotherapy and chemotherapy[5,11]. Prognosis of SNTCS is poor. Heffner and Hyams state that average survival is 1.7 years with only 40% of the patients surviving beyond 3 years[2]. Sharma et al. reports that the rate of local recurrence after surgery is approximately 43%[12]. We have reported these two cases for their rarity and would like to emphasis the need for through sampling of the tissue for arriving to the correct diagnosis.

CONCLUSION

SNTCS is a rare and aggressive tumor of sinonasal region. Good clinical diagnosis and extensive sampling is essential in these cases to avoid erroneous diagnosis. Early diagnosis, appropriate management and aggressive follow up can give better prognosis in this rare tumor.

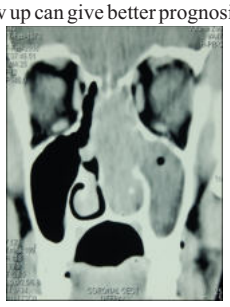

Figure 1: Computed tomography scan: non-enhancing polypoidal masses in left nasal cavity extending up to soft palate & hypopharynx

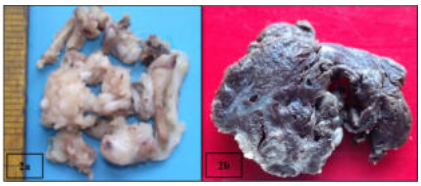

Figure 2a: Gross - Multiple pieces of whitish firm mass
Figure 2b: Gross- Reddish friable hemorrhagic mass

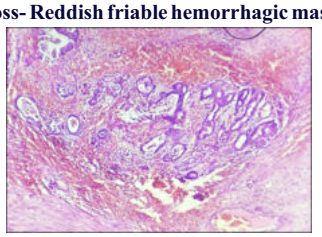

Figure 3: Photomicrograph of Glandular Component (H & E stain x 100).

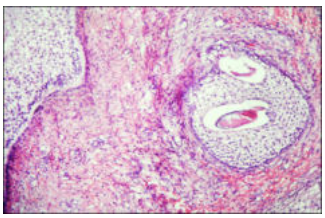

Figure 4: Photomicrograph of Immature Squamous Epithelium (H&E stain, x 400)

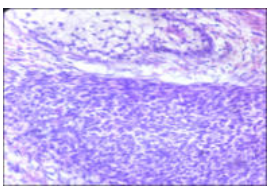

Figure 5: Photomicrograph of Neuroepithelial Components (H&E stain, x400)

REFERENCES

1. Shanmugaratnam K, Kunaratnam N, Chia KB, Chiang GS, Sinniah R. Teratoid carcinosarcoma of the paranasal sinuses. Pathology. 1983;15(4):413-9.
2. Heffer DK, Hyams VJ. Teratocarcinosarcoma of the nasal cavity and paranasal sinuses. A clinicopathological study of 20 cases. Cancer 1984; 53:2140-54.
3. Rao YF, Cheng DN, Qiu K, Song Y, Zhao Y, Gu D, Ren J. Sinonasal teratocarcinosarcoma: a case report and literature review. J Int Med Res. 2020 Dec;48(12):300060520971488. doi: 10.1177/0300060520971488. PMID: 33342340; PMCID: PMC7754799.
4. Chakraborty S, Chowdhury AR, Bandyopadhyay G. Sinonasal teratocarcinosarcoma: case report of an unusual neoplasm. J Oral Maxillofac Pathol. 2016;20(1):147-50.
5. Fatima SS, Minhas K, Din NU, Fatima S, Ahmed A, Ahmad Z. Sinonasal teratocarcinosarcoma: a clinicopathologic and immunohistochemical study of 6 cases. Ann Diagn Pathol 2013; 17(4):313–8.doi: 10.1016/j.anndiagpath.2013.01.003. Epub 2013 Feb 23.
6. Rotenberg B, El-Hakim H, Lodha A, MacCormick A, Ngan BY, Forte V. Nasopharyngeal teratocarcinosarcoma. Int J Pediatr Otorhinolaryngol [Internet]. 2002 Feb 1 [cited 2016 Nov 30];62(2):159–64.
7. Takasaki K, Sakihama N, Takahashi H. A case with sinonasal teratocarcinosarcoma in the nasal cavity and ethmoid sinus. Eur Arch Otorhinolaryngol [Internet]. 2006 Jun [cited 2016 Nov 30];263(6):586–91.
8. Pai SA, Naresh KN, Masih K, Ramarao C, Borges AM. Teratocarcinosarcoma of the paranasal sinuses: a clinicopathologic and immunohistochemical study. Hum Pathol [Internet]. 1998 Jul [cited 2016 Nov 30];29(7):718–22.
9. Fernández PL, Cardesa A, Alós L, Pinto J, Traserra J. Sinonasal teratocarcinosarcoma: An unusual neoplasm. Pathol Res Pract. 1995;191(2):166-71.
10. Shindo ML, Stanley RB, Kiyabu MT. Carcinosarcoma of the nasal cavity and paranasal sinuses. Head Neck [Internet]. [cited 2016 Nov 30];12(6):516–9.
11. Kane S V, Karpate AA, Bal M, Juvekar SL, Pai PS. Chemotherapy-induced neuronal maturation in sinonasal teratocarcinosarcoma--a unique observation. Head Neck Pathol. 2009; 3:31–6.doi: 10.1007/s12105-008-0094-x. Epub 2008 Dec 4.
12. Sharma HS, Abdullah JM, Ottoman NH, Muhamad M (1998) Teratocarcinosarcoma of the paranasal sinus: a clinicopathologic and immunohistochemical study. Hum Pathol 29:718-722.