Sinonasal teratocarcinosarcoma - A rare case report

Lopa Mudra Kakoti, Jagannath Dev Sharma, Shiraj Ahmed, Anupam Sarma

From Department of Pathology, Dr. Bhubaneswar Borooah Cancer Institute, Birubari, Guwahati, Assam, India

Correspondence to: Dr. Lopa Mudra Kakoti, Department of Pathology, Dr. Bhubaneswar Borooah Cancer Institute, Gopinath Nagar, Guwahati - 781 016, Assam, India. E-mail: lopamudrakakoti@gmail.com

Received - 04 June 2017 Initial Review - 04 July 2017 Published Online - 28 July 2017

ABSTRACT

Sinonasal teratocarcinosarcoma is a rare and highly malignant tumor characterized by complex admixture of teratomatous and carcinosarcomatous component. Here, we report a case of 72-year-old male presented with complaints of epistaxis, recurrent sinusitis, nasal stuffiness, and nasal blockade for past 5-6 months and his computed tomography report revealed as an inflammatory sinonasal disease with polyposis. Histopathology examination showed fragments of upper respiratory mucosa admixed with fragments of tumor composed of benign and malignant counterparts of both epithelial and mesenchymal components in a variety of histologic patterns. Also noted were the characteristic squamous epithelium with "fetal-type" clear cells, areas of necrosis, mitotic figure and sheets of undifferentiated blastemal like cells. Immunohistochemistry demonstrated positive staining for cytokeratin, epithelial membrane antigen, vimentin, synaptophysin, S-100, chromogranin, and CD99. This polyimmunophenotypic characteristic and diverse histological differentiation provided the basis for the diagnosis. Knowing its highly aggressive behavior, the patient should be treated aggressively and followed up closely.

Key words: *Carcinosarcom, Fetal epithelium, Polyimmunophenotypic, Teratoid*

Sinonasal teratocarcinosarcoma is a rare aggressive malignant tumor characterized by complex admixture of teratomatous and carcinosarcomatous component [1]. In literature, different names have been documented such as blastemal, malignant teratoma, and teratocarcinoma. First reported case was by Shanmugaratnam et al. under the term "teratoid carcinosarcoma" [2]. Later in 1984, Heffner and Hyams reported a series of similar cases modifying the term as "teratocarcinosarcoma" [3]. Till now, around 66 cases have been documented [4]. The lesion is often misdiagnosed due to rarity and the complex phenotypic composition and inadequate sampling. We present a rare case report from North-East India along with related review of literature of the same.

CASE REPORT

A 72-year-old male presented with complaints of epistaxis, recurrent sinusitis, nasal stuffiness, and nasal blockade for past 5-6 months and was being treated in an outside private clinic. There was no significant past medical or family history (non-tobacco user), and his physical examination did not reveal any abnormal systemic features. The clinical diagnosis was made as allergic polyp based on endoscopic findings of multiple tiny smooth gray white glistening polypoidal masses. His computed tomography scan also showed inflammatory sinonasal disease with polyposis at places including left sided nasal choanal polyp and his routine investigations were within normal limit. The patient had undergone polypectomy at outside clinic and biopsy was sent to our hospital.

The specimen was sent in 3 containers and grossly consists of multiple fragmented bits of tissue collectively amounting from 2.5 × 2.5 × 2 cm^3 to 1.5 cm across, gray white to gray brown in color. Histopathology sections showed fragments of the upper respiratory mucosa lined by ciliated columnar epithelium admixed with fragments of tumor composed of benign and malignant counterparts of both epithelial and mesenchymal components in a variety of histologic patterns. Also noted were the characteristic squamous epithelium with "fetal-type" clear cells (Fig. 1), areas of necrosis and increased mitotic figure. There were sheets of densely-packed small cells with scanty cytoplasm and uniform rounded nuclei resembling undifferentiated blastemal cells with proliferative neuroepithelium in the form of tubules. This primitive neural tissue was surrounded by myxoid area (Fig. 2) with elongated or rounded nuclei with intervening bundles of fibroblastic cells (Fig. 3).

Immunohistochemistry (IHC) demonstrated positive staining for cytokeratin, epithelial membrane antigen, vimentin, synaptophysin, S-100, chromogranin, GFAP, and CD99 (Figs. 4-6). This poly immunophenotypic characteristic and diverse histological differentiation provided the basis for the diagnosis of sinonasal teratocarcinoma. Further, the patient was referred for radiation therapy, and he was responding well. The patient has been closely followed up without any complaints for last 6 months.

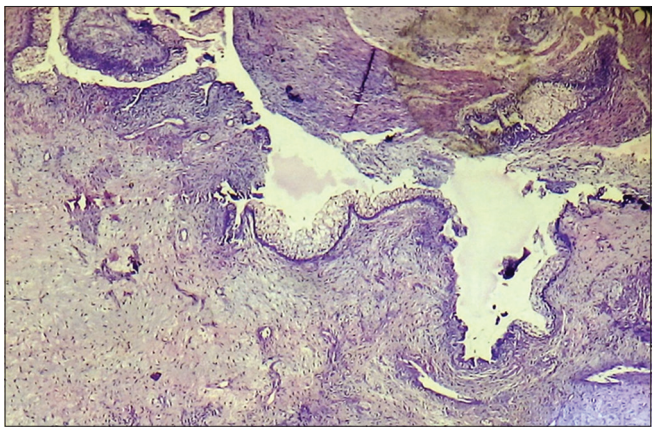

Figure 1: H and E, ×20, squamous epithelium with "fetal-type" clear cells and myxoid area

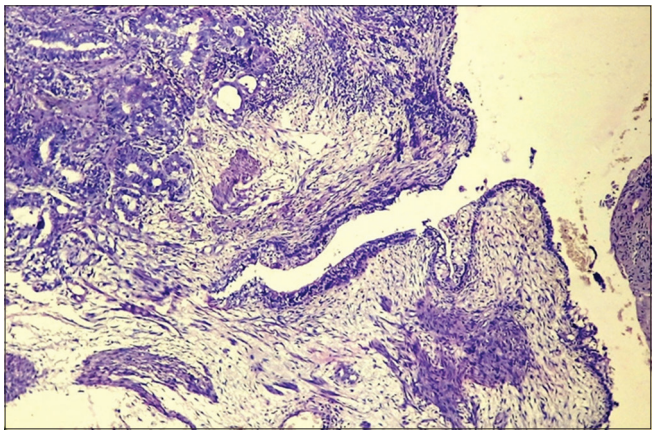

Figure 2: H and E, ×20, neural tubules and blastemal cells

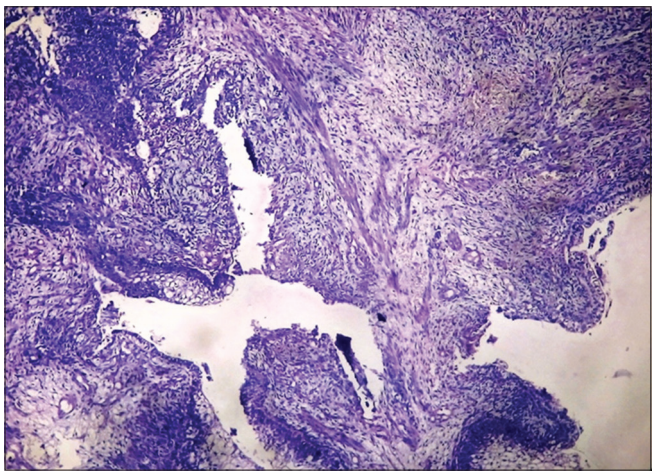

Figure 3: H and E, ×20, bundles of fibroblastic cells

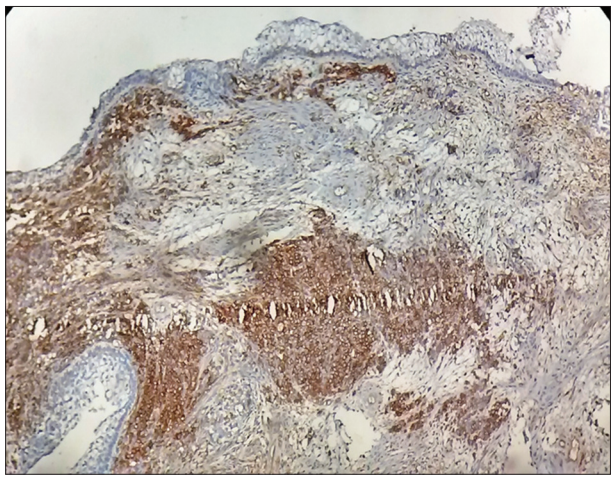

Figure 4: Immunohistochemistry, ×20, CD99 positivity in blastemal cells

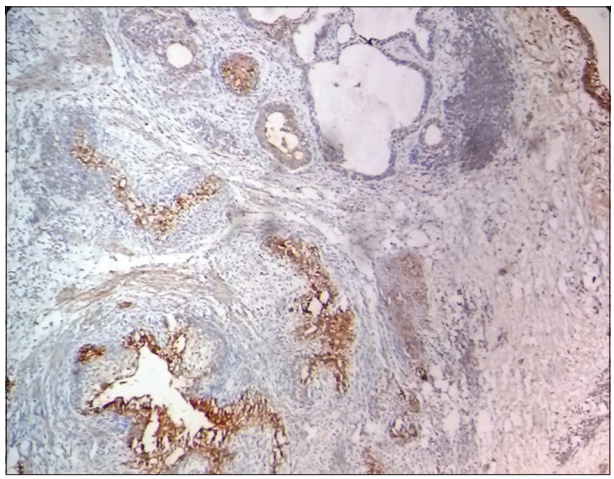

Figure 5: Immunohistochemistry, ×20, S100 positivity in neural tubular epithelium

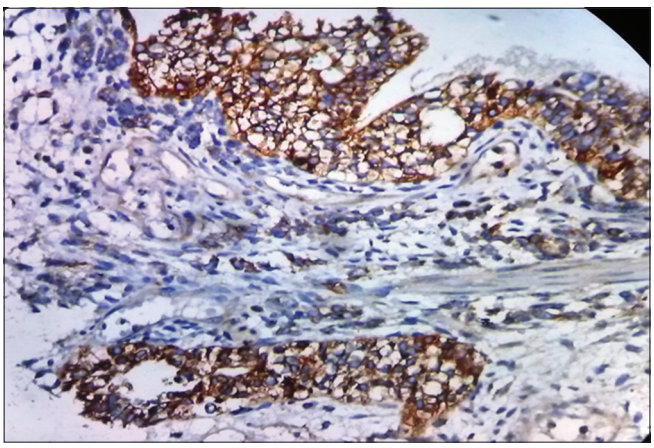

Figure 6: Immunohistochemistry, ×40, cytokeratin positive fetal epithelium

DISCUSSION

Teratocarcinosarcoma, a rare highly malignant polymorphous neoplasm that displays combined features of an immature or malignant teratoma and a carcinosarcoma. It often tends to occur in the nose, pharynx and sinus areas, but tumor in other parts of the body has also been recorded. Clinical features are determined by the size and location of the tumor. The most common complaints at presentation include nasal obstruction, epistaxis, facial pain, headache, proptosis, and visual field deficits [2]. This neoplasm is more commonly seen in male with male to female ratio 4:1 and between 18 and 79 years of age [2]. Our patient was 72-year-old male presented with most common presentation such as epistaxis and nasal blockade.

Teratocarcinosarcoma is highly aggressive and locally recurrent tumor. Less is known about its metastatic potential

except very few reports mentioning about their metastatic potential to regional lymph nodes as well as in the lungs [5]. Hence, this very aggressive tumor requires multidisciplinary approach. Surgical resection with post-operative radiotherapy being the widely accepted therapeutic plan [5,6]. Role of chemotherapy is controversial; however, Sharma et al. have reported effective control of tumor recurrence under control (for a short period) with adjuvant chemotherapy [6]. Good responses were showed when adjuvant multidrug chemotherapy with cisplatin, etoposide, and ifosfamide was used [7].

Recently, Joshi et al. 2015 [8] have reported a recurrent case of sinonasal carcinosarcoma which was inoperable because of its cranial extension, and he was treated with palliative chemotherapy with ifosfamide and doxorubicin, and the patient showed significant symptomatic relief. Despite aggressive treatment, the prognosis is poor. The local recurrence after excision was found to be high with reported 3 and 5 years survival rates of about 30% and 20%, respectively [8]. Our patient was given radiotherapy, and he was responding well. For the last 6 months, he has been closely followed up without any complaints. Differential diagnosis includes poorly differentiated carcinoma, sarcoma, and olfactory neuroblastoma. IHC helps to rule out the differential diagnosis.

CONCLUSION

Sinonasal teratocarcinosarcoma is a rare and highly malignant tumor. “Fetal-type” clear cells squamous epithelium and immature neuroepithelium represent an important histologic clue to the histomorphological diagnosis and poly immunophenotypic characteristic in IHC provides the basis for its diagnosis. Knowing its highly aggressive behavior, the patient should be closely followed up, and chemotherapy can be integrated as an adjuvant or palliative setting.

REFERENCES

1. Fernández PL, Cardesa A, Alós L, Pinto J, Traserra J. Sinonasal teratocarcinosarcoma: An unusual neoplasm. Pathol Res Pract. 1995;191(2):166-71.
2. Shanmugaratnam K, Kunaratnam N, Chia KB, Chiang GS, Sinniah R. Teratoid carcinosarcoma of the paranasal sinuses. Pathology. 1983;15(4):413-9.
3. Heffner DK, Hyams VJ. Teratocarcinosarcoma (malignant teratoma?) of the nasal cavity and paranasal sinuses a clinicopathologic study of 20 cases. Cancer. 1984;53(10):2140-54.
4. Wahid FI, Javaid M, Khan Q, Khan IA. Sinonasal teratocarcinosarcoma. J Coll Physicians Surg Pak. 2012;22(5):335-7.
5. Kazemian A, Manshadi HR, Kermani S. Sinonasal teratocarcinosarcoma; report of a case. Iran J Radiat Res. 2004;2:159-62.
6. Sharma HS, Abdullah JM, Othman NH, Muhamad M. Teratocarcinosarcoma of the nasal cavity and ethmoid. J Laryngol Otol. 1998;112(7):682-6.
7. Nitsche M, Hermann RM, Christiansen H, Berger J, Pradier O. Rationale for individualized therapy in sinonasal teratocarcinosarcoma (SNTC): Case report. Onkologie. 2005;28(12):653-6.
8. Joshi A, Dhumal SB, Manickam DR, Noronha V, Bal M, Patil VM, et al. Recurrent sinonasal teratocarcinosarcoma with intracranial extension: Case report. Indian J Cancer. 2014;51(3):398-400.

Funding: None; Conflict of Interest: None Stated.

How to cite this article: Kakoti LM, Sharma JD, Ahmed S, Sarma A. Sinonasal teratocarcinosarcoma - A rare case report. Indian J Case Reports. 2017;3(3):164-166.