

Case report

Fibromyxoid sarcoma of maxilla

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ABSTRACT

A recurrent mass in the nose can pose diagnostic and therapeutic challenges. Even more so, if it exhibits rapid growth, displays unusual clinical features, is a rare histopathological diagnosis or if the patient is unavailable for regular follow-up. We present a case of a middle-aged retrovirus-positive man who underwent surgical removal of a recurrent nasal mass. Histopathology revealed a diagnosis of low-grade fibromyxoid sarcoma, a rare entity in the sinonasal tract. Immunohistochemistry plays a crucial role in clinching the diagnosis, especially for lesions with fibrous and myxoid components. Early and complete surgical resection diminishes the likelihood of recurrence and metastasis.

Keywords: Maxilla; immunohistochemistry; nasal neoplasms; sarcoma; sinonasal tract.

INTRODUCTION

Most sinonasal masses present alike. Probable diagnosis may be made with imaging (1) confirmed by biopsy; endoscopic management is sufficient in most instances. Nonetheless, recurrent lesions pose a challenge in management, especially when growth is rapid, clinical features are peculiar, and if the patient is unavailable for regular follow-up.

CASE REPORT

A 47-year-old gentleman, known case of Human Immunodeficiency Virus (HIV) infection (CD4 count 306 and HIV-1 viral load <20 copies/ml), on anti-retroviral treatment, presented with left nasal obstruction for two months. Examination revealed a polypoidal mass arising from the left lateral nasal wall, completely occluding the left nasal cavity (Fig. 1).



Fig. 1: Endoscopic photograph showing a pale mass in the left nasal cavity (asterisk), 5mm behind nasal vestibule, completely occluding left nasal cavity.

Contrast-enhanced computed tomography (CECT) findings revealed an ill-defined iso-dense mass with thick peripheral enhancement in the left nasal cavity

and maxillary sinus. With a clinical suspicion of inverted papilloma, left endoscopic medial maxillectomy was performed with complete clearance. The possibility of Kaposi sarcoma or Non-Hodgkin's Lymphoma (NHL) was also contemplated clinically owing to the patient's retrovirus-positive status. However, histopathology was suggestive of spindle cell neoplasm; immunohistochemistry (IHC) showed focally positive smooth muscle actin (SMA), diffusely positive cytokeratin, 8% Ki-67, negative CD34, human melanoma black 45, S-100, anaplastic lymphoma receptor tyrosine kinase and desmin, suggestive of pseudosarcomatous myofibroblastic proliferation.

The patient was informed about the condition and advised of periodic follow-up and the need for revision surgery in the event of recurrence. However, he visited the hospital only two months later; the swelling had returned and repeat CECT (Fig. 2) revealed erosion of the posterior maxillary wall.

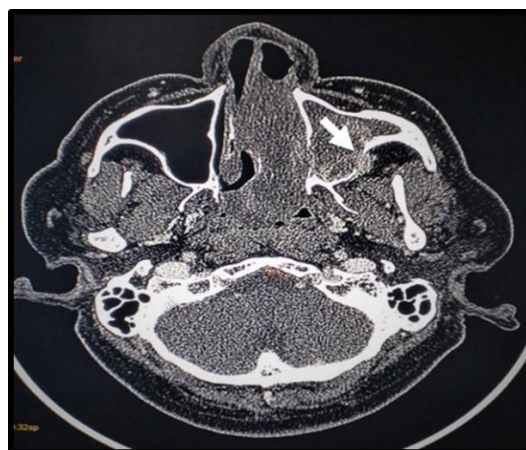


Fig. 2: Contrast-enhanced computed tomography (CECT) image revealing erosion of left posterior maxillary wall (white arrow).

Left total maxillectomy was performed. Histopathological examination showed spindle cell proliferation in sheets, fascicles and myxoid appearance, (Fig. 3A) moderately pleomorphic

tumour cells with spindle-shaped hyperchromatic nuclei, small nucleoli and a moderate amount of tapering cytoplasm.

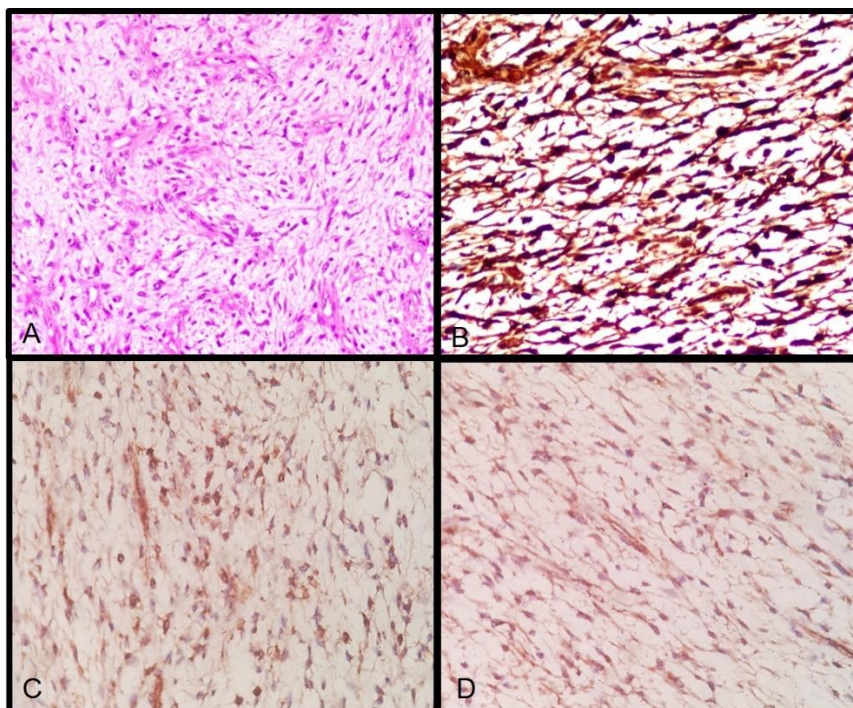


Fig. 3: **A.** Photomicrograph of low-grade fibromyxoid sarcoma (LGFMS) with proliferating spindle cells in fascicles having myxoid appearance (200x, H&E) **B.** Photomicrograph of immunohistochemistry (IHC) pattern in LGFMS with strong positivity for vimentin (200x) **C.** Positivity for CD99 (100x) **D.** Tumour cells showing cytoplasmic positivity for beta-catenin and absence of nuclear positivity on immunohistochemistry (100x).

Interspersed were many thin-walled blood vessels; stroma was predominantly myxoid to focally collagenous. Infrequent areas of necrosis and atypical mitosis were noted. Immunohistochemistry showed strong positivity for vimentin (Fig. 3B), positivity for CD99 (Fig. 3C) and weak positivity for SMA. The tumour cells showed cytoplasmic positivity for beta-catenin and absence of nuclear positivity (Fig. 3D), negative for Bcl-2, p40 and epithelial membrane antigen. All margins except lateral were free of tumour. Clinicoradiopathological and IHC features were correlated, and a diagnosis of low-grade fibromyxoid sarcoma (LGFMS) was considered. The patient was referred to a higher centre for mucin-4 (MUC-4) marker and molecular testing, as they were not offered at our hospital. However, he was lost to follow up. His relatives confirmed that he had not considered any further management and had expired about six months after maxillectomy.

DISCUSSION

Our patient presented with unilateral nasal obstruction and a left-sided nasal mass; clinically, inverted papilloma was regarded the first diagnosis, as it is known to occur as a polypoidal mass, has a high recurrence rate, and causes bone erosion (2). A diagnosis of Kaposi sarcoma or NHL could not be ruled out clinically. While external approaches were once exclusively performed for most of the sinonasal tumours, endoscopic clearance has attained

acceptance due to reduced morbidity (2). Since the mass was limited, endoscopic medial maxillectomy was performed on our patient.

Unpredictably, histopathological diagnosis favoured pseudosarcomatous myofibroblastic proliferation (PMP), an unusual entity, also considered inflammatory myofibroblastic tumour (IMT), with malignant potential. The lack of typical clinico-radiological characteristics makes preoperative diagnosis difficult (3). It is commoner in the genitourinary tract than the head and neck (submandibular space, maxillary region, oral cavity; 3,4). Steroids and methotrexate have been used with surgery for the successful management of maxillary IMT (5). Unfortunately, they were not considered in our patient owing to his immunocompromised state.

The final diagnosis after total maxillectomy was the most appalling. Head and neck sarcomas represent 10% of head and neck tumours and 4-10% of all soft tissue sarcomas; sinonasal occurrence is even rarer (6,7). Ewing's sarcoma is the commonest subtype (28.3%), followed by malignant fibrous histiocytoma (12.6%). Maxillary sinus and nasal cavity are frequent sites of sinonasal sarcoma (58.1% and 55.6%, respectively); five-year overall survival is 40-90% (6).

LGFMS is a rare sarcoma with an indolent clinical course affecting the younger population (8). Only a handful of cases have been reported in the head and neck, predominantly mandible and larynx. Due to its

rarity, pathologists may not consider LGFMS as a differential; immunohistochemistry is valuable. MUC-4 positivity and molecular testing are confirmatory (9). The unavailability of these tests in our hospital led to a higher centre referral. The patient's unwillingness for further evaluation and the onset of the coronavirus pandemic contributed to his early demise. Wide surgical resection with clear margins is the standard management of LGFMS. It is unlikely to respond to chemoradiation as it is a low-grade malignancy with a low mitotic rate. Recurrence is 10%, and late metastasis rate is 5% (8).

CONCLUSION

LGFMS is rare in the sinonasal region; it has a prolonged subclinical presentation. IHC plays a crucial role in clinching the diagnosis, especially for lesions with fibrous and myxoid components; this entity must be kept in mind in such cases, enabling early, adequate surgical resection and diminishing the likelihood of recurrence and metastasis.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

REFERENCES

1. Agarwal, P., Panigrahi, R. Sinonasal Mass — a Recent Study of Its Clinicopathological Profile. *Ind J Surg Oncol.* 2017; 8: 123-127. <https://doi.org/10.1007/s13193-016-0570-9>.
2. Vorasubin, N., Vira, D., Suh, J. D., Bhuta, S., Wang, M. B. Schneiderian papillomas: Comparative review of exophytic, oncocytic, and inverted types. *Am J Rhinol Allergy.* 2013; 27: 287-292. <https://doi.org/10.2500/ajra.2013.27.3904>.
3. Tanaka, T., Ueda, T., Yokoyama, T., Harada, S., Hatakeyama, K., Yoshimura, A. Pseudosarcomatous myofibroblastic proliferation of the appendix with an abdominal abscess due to diverticulum perforation: a case report. *Surg Case Reports* 2020; 6: 144. <https://doi.org/10.1186/s40792-020-00901-1>.
4. Dosemane, D., Khadilkar, U., Khadilkar, M. Seek the Spindle Tumor in Submandibular Space. *Iran J Otorhinolaryngol* 2019; 31: 181-184. <https://doi.org/10.22038/IJORL.2019.28427.1931>.
5. Chong, S., The, C.S.L., Singh, S., Seong, M. K., Viswaraja, S. Aggressive inflammatory pseudotumor of the maxillary sinus. *Ear Nose Throat J* 2014; 93: 108-111. <https://doi.org/10.1177/014556131409300308>.
6. Gore, M. R. Treatment, outcomes, and demographics in sinonasal sarcoma: a systematic review of the literature. *BMC Ear Nose Throat Disord.* 2018; 18: 1-13.
7. Stavarakas, M., Nixon, I., Andi, K., Oakley, R., Jeannon, J. P., Lyons, A., *et al.*, Head and neck sarcomas: Clinical and histopathological presentation, treatment modalities, and outcomes. *J Laryngol Otol.* 2016; 130: 850-859. <https://doi.org/10.1017/S0022215116008604>.
8. Sambri, A., Righi, A., Tuzzato, G., Donati, D., Bianchi, G. Low-grade fibromyxoid sarcoma of the extremities: A clinicopathologic study of 24 cases and review of the literature. *Pol J Pathol* 2018; 69: 219-225. <https://doi.org/httpS://doi.org/10.5114/pjp.2018.79541/>.
9. Cowan, M. L., Thompson, L. D., Leon, M. E., Bishop, J. A. Low-Grade Fibromyxoid Sarcoma of the Head and Neck : A Clinicopathologic Series and Review of the Literature. *Head Neck Pathol* 2016; 10: 161-166. <https://doi.org/10.1007/s12105-015-0647-8>.