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Case Report

Navigating the Uncommon: Rare Cases in Head and Neck Oncology

Girish Mishra[1], Simran J Parmar[2]

[1-2]Department of ENT and Head and Neck Surgery, pramukhswami medical college and Bhaikaka University karamsad, Anand, Gujarat1, Simran J Parmar2

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Corresponding Author:

Girish Mishra
Department of ENT and Head and Neck Surgery, pramukhswami medical college and Bhaikaka University karamsad, Anand, Gujarat1, Simran J Parmar2

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ABSTRACT

Head and neck cancers encompass a broad spectrum of neoplasms, ranging from benign to highly aggressive malignancies. Accurate diagnosis and timely management are crucial to improving outcomes. This case series presents five rare and histopathologically diverse head and neck tumours like mammary analogue secretory carcinoma, paraganglioma, adenoid cystic carcinoma, esthesioneuroblastoma and cutaneous squamous cell carcinoma. Each case highlights unique clinical presentations, radiological findings, and surgical interventions. The histopathological evaluation served as the gold standard in establishing definitive diagnoses. Early recognition and a multidisciplinary approach remain vital for optimising prognosis in these uncommon entities.

Keywords*: Head and neck cancer, rare tumours, histopathology, parotid carcinoma, esthesioneuroblastoma, squamous cell carcinoma.*

INTRODUCTION

Head and neck oncology represents one of the most anatomically and biologically diverse fields in cancer medicine, encompassing malignancies that arise from the upper aerodigestive tract, salivary glands, and skin[1]. Despite substantial advances in surgical precision, imaging and radiotherapy, early diagnosis remains the cornerstone for improved survival2. Among these neoplasms, rare histopathological variants such as mammary analogue secretory carcinoma (MASC), paraganglioma, adenoid cystic carcinoma (ACC), esthesioneuroblastoma (ENB), and cutaneous squamous cell carcinoma (SCC) of unusual sites challenge clinicians due to their overlapping clinical presentations and unpredictable biological behavior.3,4 The subtle clinical features often mimicking benign or inflammatory lesions frequently delay diagnosis, leading to advanced-stage detection. Histopathological evaluation, supplemented by immunohistochemistry and molecular profiling, provides the definitive diagnosis. Recent discoveries, such as the ETV6-NTRK3 gene fusion in MASC and the identification of specific neuroendocrine markers in ENB have revolutionised diagnostic accuracy and targeted therapy options5,6. This case series highlights five rare and histopathologically distinct tumours encountered at a tertiary care centre. Through detailed clinicopathologic and surgical correlation aims to enhance understanding of diagnostic nuances and emphasize the need for multidisciplinary management in improving prognosis.

Aim: To investigate and analyze rare cases of head and neck cancer, with a focus on understanding their incidence, clinical characteristics, treatment outcomes, and histopathological correlation.

CASE PRESENTATION

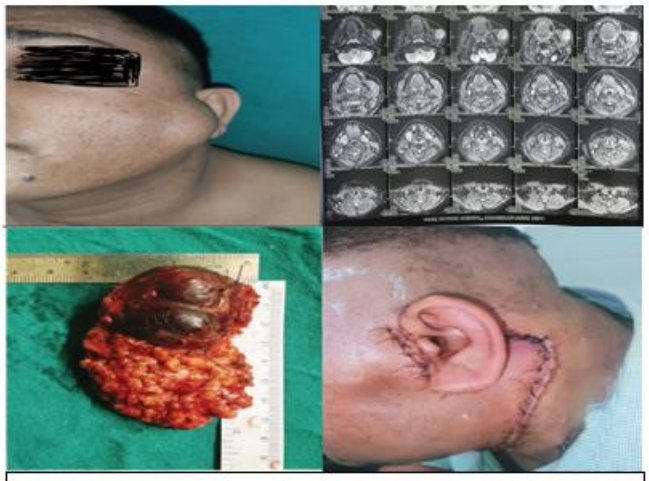

Figure 1: Clinical picture of left parotid swelling, MRI image, Intraop picture of left superficial parotidectomy, post operative photograph

Case 1: Mammary Analogue Secretory Carcinoma / Mucoepidermoid Carcinoma of Parotid Gland

- A 24-year-old male presented with swelling over the left parotid region for 6 months.
- Imaging: Ultrasound showed a well-defined cystic lesion with no internal vascularity.
- MRI showed Well defined solid-cystic lesion of size 3.9 x 3.5 x 3.3 cm with central solid component noted in left parotid region.The lesion is infiltrating left parotid gland and left masseter muscles. Multiple subcentimetric to enlarged enhancing lymphnodes are noted in cervical level IA, bilateral IB, bilateral IIA, bilateral IIB, III and bilateral Vb region.
- FNAC: Category IVa, benign lesion of the left parotid gland.
- Surgery: Left superficial parotidectomy was performed.

Histopathology: Revealed mammary analogue secretory carcinoma with areas of mucoepidermoid differentiation. This highlights the role of histopathology in differentiating rare parotid tumors with overlapping cytological features.

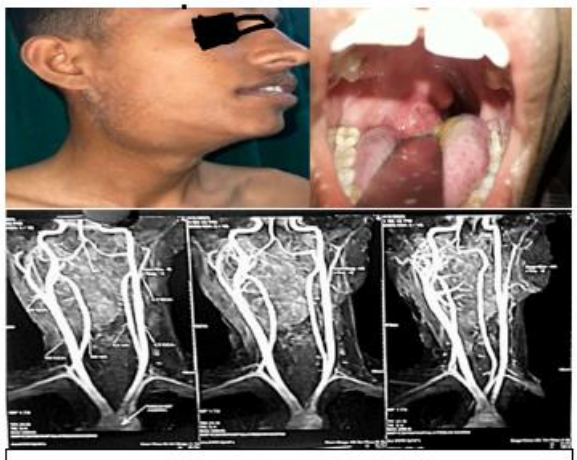

Figure 2: showing clinical photograph of right submandibular swelling and right tonsillar region, MR angiogram photograph,

Case 2: Paraganglioma /Alveolar Soft Part Sarcoma A 16-year old male presented with dysphagia and a swelling over the right submandibular and tonsillar regions for 8 months.

Surgery: Right transcervical excision of the mass with reconstruction using the sternal head of the sternocleidomastoid muscle.

Histopathology: Revealed features consistent with paraganglioma and focal alveolar soft part sarcoma. The dual pathology made the case diagnostically challenging and highlights the value of immunohistochemistry in differentiating overlapping entities.

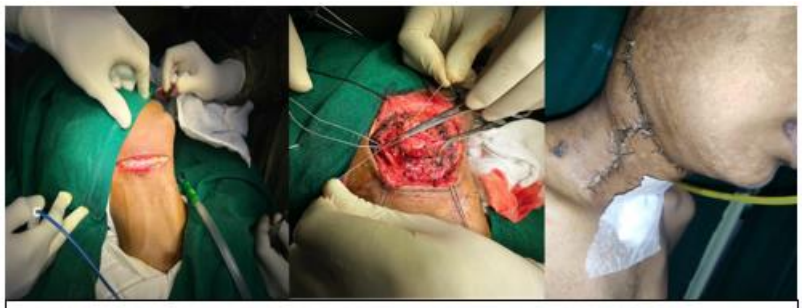

Figure 3: Intraoperative and post operative photograph

Case 3: Adenoid Cystic Carcinoma of Nasal Cavity A 45 year old female presented with right-sided nasal obstruction for 1 year and a protruding nasal mass for 4 months.

CT Scan showed a polypoidal lesion involving the middle and inferior meatus with bony erosion.

Surgery: Endoscopic excision of the mass was done Histopathology: Confirmed adenoid cystic carcinoma. This case underscores the infiltrative nature of ACC and the importance of achieving clear surgical margins.

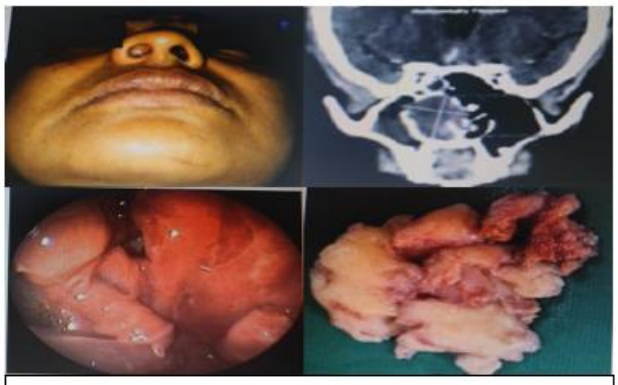

Figure 4: Clinical photograph of right nasal mass, CT image, Intra operative photograph and Specimen after excision

Case 4: Esthesioneuroblastoma (Olfactory Neuroblastoma)

- A 16-year-old male presented with right nasal blockage and right eye proptosis with chemosis for 3 months.
- The patient had previously undergone endoscopic sinus surgery elsewhere, where histopathology suggested esthesioneuroblastoma.
- MRI: Showed a lesion involving the right rhino-orbital region with bony erosions along the olfactory fossa.
- Surgery: Revision endoscopic sinus surgery with orbital decompression and tumor debulking was done
- Adjuvant therapy: 33 cycles of radiotherapy and 5 cycles of chemotherapy. Outcome: Postoperative PET scan was normal, and proptosis resolved completely.

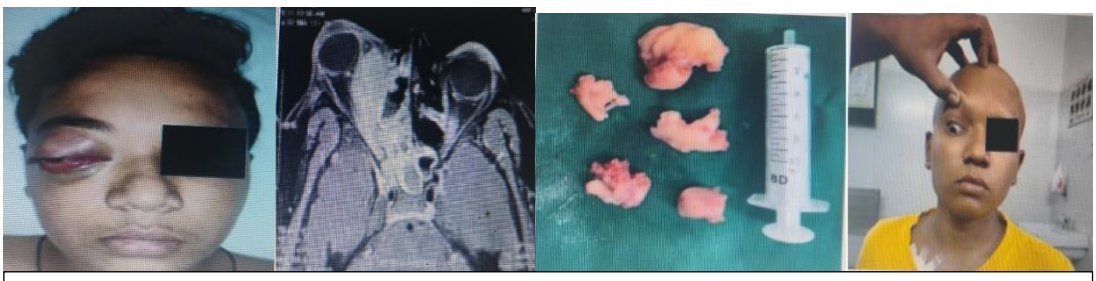

Figure 5: Clinical photograph with right eye swelling, MRI image, Specimen after tumour debulking, post operative photograph

Case 5: Cutaneous Squamous Cell Carcinoma of Nose A 44-year-old female presented with a non-healing ulcer over the nose for 2 months.

Biopsy: Moderately differentiated squamous cell carcinoma. CT Scan: Showed exophytic soft-tissue lesion at the nasal tip. Surgery: Wide excision with forehead flap reconstruction and split-thickness skin grafting done from the thigh. Histopathology: Confirmed cutaneous SCC (pT2).

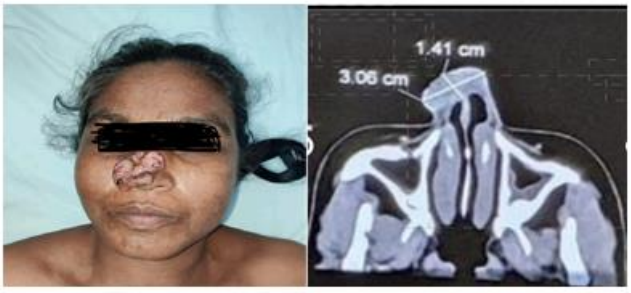

Figure 6: Clinical photograph, CT image

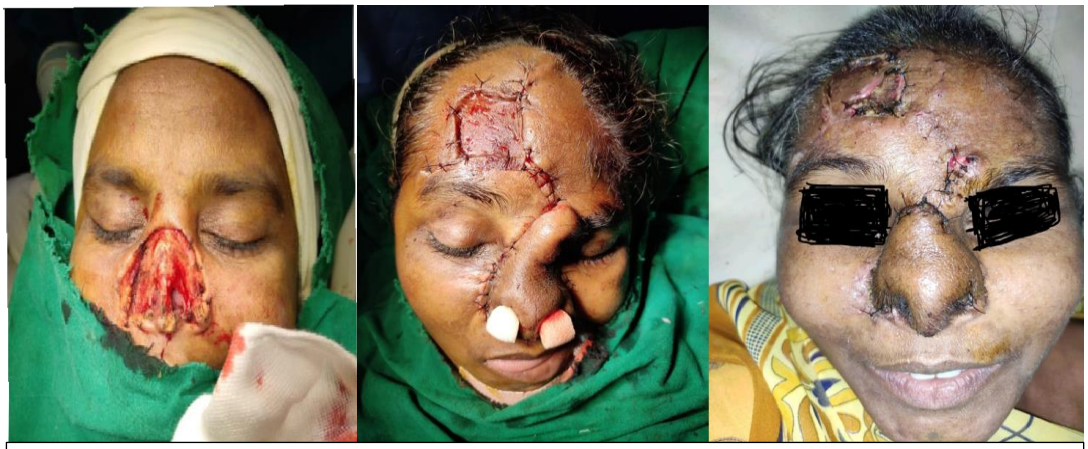

Figure 7: Intra-operative and post operative photographs

Early surgical intervention with complete excision provided an excellent cosmetic and functional outcome.

DISCUSSION

Head and neck tumours comprise an exceptionally heterogeneous group of diseases differing in embryological origin, histogenesis and clinical behaviour. The management of rare variants requires a comprehensive integration of clinical acumen, radiological interpretation, and histopathological expertise.

Mammary analogue secretory carcinoma (MASC), first characterised in 2010, is an uncommon salivary gland malignancy that shares morphological and molecular resemblance with secretory breast carcinoma. The hallmark ETV6-NTRK3 translocation, identifiable through fluorescence in situ hybridisation (FISH) has therapeutic implications with the emergence of TRK inhibitors such as larotrectinib and entrectinib8. Distinguishing MASC from acinic cell carcinoma is essential, as the latter lacks this molecular marker.

Paragangliomas of the head and neck region exhibit indolent yet locally invasive growth. Genetic testing for SDHB and SDHD mutations is increasingly advocated due to their association with familial syndromes and metastatic potential9. The unusual co-existence of paraganglioma and alveolar soft part sarcoma, as seen in our series, underscores the diagnostic complexity of these lesions.

Adenoid cystic carcinoma (ACC) is well recognised for its perineurial invasion and late distant metastasis. Studies have shown that surgical resection with negative margins followed by adjuvant radiotherapy remains the gold standard10. Long-term surveillance is crucial, as recurrences may occur even after a decade11.

Esthesioneuroblastoma(ENB) arising from olfactory neuroepithelium, represents 3-6% of all sinonasal malignancies12. Kadish staging continues to guide management, while Hyams histological grading predicts prognosis. Combined modality treatment with surgery, radiotherapy and chemotherapy offers the best outcomes, particularly in locally advanced disease13.

Cutaneous squamous cell carcinoma (SCC) of the nasal tip poses both oncologic and reconstructive challenges. Early stage lesions (T1&T2) have excellent survival when treated with wide local excision and flap reconstruction. Delayed diagnosis or incomplete resection, however, can lead to local recurrence and cosmetic deformity14.

Across all these rare tumors, the integration of histopathology, molecular profiling, and multidisciplinary tumor boards ensures optimal management. Furthermore, emerging research on targeted therapies including NTRK inhibitors for MASC, tyrosine kinase inhibitors for paragangliomas, and proton therapy for sinonasal malignancies has transformed outcomes15,16.

Despite progress, the rarity of these entities limits large-scale clinical trials. Therefore, case documentation and multicenter registries remain invaluable in refining diagnostic algorithms and treatment protocols17.

CONCLUSION

This series reinforces that histopathological assessment remains the gold standard for definitive diagnosis in head and neck oncology. Early identification, adequate surgical margins, and multimodal treatment significantly improve survival rates. Continued reporting of such rare entities contributes to building a robust evidence base, facilitating earlier recognition, and improving patient care outcomes.

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