Diverse Diagnosis of a Single Sinonasal Mass and how was Mystery Solved
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Abstract

Background: Although the nasal cavity and paranasal sinuses occupy a relatively small anatomical space, they are the site of origin of some of the more complex, histologically diverse group of tumours in the entire human body. Carcinomas of the nasal cavity and paranasal sinuses account for 0.2-0.8% of all malignant neoplasms and 3% of those occurring in the head and neck. Sinonasal undifferentiated carcinoma is a very rare tumour with fewer than 100 reported cases.

Case report: In our case, a 50yr female came to the ENT OPD with a mass in the right nasal cavity with respiratory distress. CECT revealed an inverted papilloma or a malignant mass lesion. Histopathological examination of the resected specimen suggested that of Transitional Cell Carcinoma (TCC). Patient again presented after 6 months, with epistaxis and right maxillary growth. A punched biopsy specimen was sent which on histopathology suggested that of an inflammatory polyp. Whole specimen was resected after that and on haematoxylin and eosin section showed the picture of Undifferentiated Carcinoma. Immunohistochemical study helped in confirmation (CK positive).

Discussion: Sinonasal undifferentiated Ca is reported as high grade malignant neoplasm of nasal cavity of uncertain histogenesis. In our case diverse histopathological diagnosis was made in single tumour as TCC (September 2014) and SNUC (April 2015, final diagnosis) when section was taken from fully resected sinonasal mass. The picture was consistent with Inflammatory Polyp (March 2015) on punched biopsy specimen was examined. So at times HPE can be misleading. However IHC is confirmatory.

Conclusion: In limited biopsy material, differentiation of these tumour types can be challenging. The histopathology of entire tumour plays a primary role in establishing the correct diagnosis which often necessitates the use of adjunct studies that allow confirm differentiation among neoplasms.

Keywords: Undifferentiated carcinoma; Sinonasal mass; IHC

Introduction

Although the nasal cavity and paranasal sinuses occupy a relatively small anatomical space, they are the site of origin of some of the more complex, histologically diverse group of tumours in the entire human body. Carcinomas of the nasal cavity and paranasal sinuses account for 0.2-0.8% of all malignant neoplasms and 3% of those occurring in the head and neck.

Sinnonasal undifferentiated carcinoma is a highly aggressive and clinicopathologically distinctive carcinoma of uncertain histogenesis that typically presents with locally extensive disease [1]. It was first reported by Frierson et al. [2]. SNUC affects more males (2-3:1) with a broad age range (third to ninth decade), and the median age is in the sixth decade.

Case History

In our case, a 50 yr old female came to the ENT OPD on Sept, 2014 with increased respiratory difficulty; discharge from the eyes and with pain in the nose and eyes. She was on herbal medicines for 1 yr (Figure 1).

Clinician on examination found a mass over right nasal cavity. CECT showed an enhancing mass lesion in the right nasal cavity and right maxillary sinus. Excision of mass was done by right lateral rhinotomy.

Gross examination showed grayish white tissue with solid areas of haemorrhage. Histopathological examination of A and B section from the tissue showed elongated cells with a cylindrical or columnar appearance, oriented perpendicular to the surface, without any evidence of keratinisation. The picture was consistent with transitional cell carcinoma (non-keratinizing squamous cell carcinoma) with areas of haemorrhage and necrosis (Figure 2).

After a period of 6 months, patient again presented with bleeding nose. Clinician on examination found features of epistaxis. Following which CECT was done, which showed heterogeneously enhancing sinonasal mass lesion involving right maxillary sinus and nasal cavity along with its regional extension. Features suggested possibility of the following –D/D- Inverted papilloma or malignant mass.

Punch biopsy was done and a small bit of tissue was sent for diagnosis. Haematoxylin and Eosin staining of the section showed only loose oedematous fibrocollagenous stromal tissue, infiltrated by chronic inflammatory cells mostly lymphocytes with extensive areas of haemorrhage. As epithelial tissue was not noted in that small bit of tissue or no evidence of malignant cell infiltration seen in the stroma, comment on inverted papilloma or malignancy could not be made. The overall picture went in favor of inflammatory polyp (Figure 3).

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Afterwards the whole mass was excised using Weber Fergocias incision under GA and specimen was sent for HPE. Gross specimen showed a soft irregular mass with brownish, irregular external surface. The mass on cut section was grayish white with few areas of hard consistency.

Microscopic picture on HandE section revealed a hypercellular neoplasm showing trabecular and lobular growth patterns. At higher magnification, cellular infiltrate contained pleomorphic cells with round to oval hyperchromatic nuclei, prominent nucleoli and increased mitotic activity. No evidence of squamous or glandular differentiation was noted. The histological picture was consistent with undifferentiated sinonasal carcinoma (Figure 4).

Immunohistochemistry was performed which showed Cytokeratin positivity (Figure 5A), while it showed negative immunoreactions for chromogranin A, LCA and S100 (Figure 5B and 5C) which established it as carcinoma.

**Discussion**

Generally, SNUC is extensive at presentation and involves multiple sites, including the nasal cavity, one or more paranasal sinuses, orbit, skull base, and the brain [2]. Our case also showed extensive involvement of the right maxillary sinus, extension into the right nasal cavity and to the right posterior ethmoidal air cells at presentation. Typically, patients present with multiple symptoms that include nasal obstruction, epistaxis, proptosis, visual disturbances (e.g., diplopia), facial pain, and symptoms of cranial nerve involvement. In our case too patient presented with symptoms of nasal obstruction, epistaxis and facial pain.

![Figure 1: Picture of the patient after teletherapy.](image1)

![Figure 2: Showing sheets of elongated cells oriented perpendicular to the surface (Transitional Carcinoma) (40X).](image2)

![Figure 3: Showing picture of inflammatory polyp (10X).](image3)

![Figure 4: Showing undifferentiated features (40X).](image4)

![Figure 5a: Showing positive staining for cytokeratin (40X).](image5a)
However, possibility of olfactory neuroblastoma (high Grade) and small cell neuroendocrine carcinoma should be excluded, given the overlapping clinical, light microscopic, immunohistochemical, and ultrastructural features with both these neoplasms [3].

Other differential diagnosis can be Lympho epithelial carcinoma (SNUC is entirely epithelial with secondary lymphoid component) or Mucosal malignant melanoma.

Although differences can be identified microscopically, but other differentiation of tumour types rests on immunohistochemical markers which shows, CK positivity in SNUC; NS positivity in ONB; CK, p63 positivity in NPC and S-100 positivity in MMM

Overall prognostic outcome of SNUC is poor. An aggressive approach using surgery, platinium chemo and radiation offer greatest chance of locoregional control and survival. Death occurs within short periods. It is an aggressive neoplasm. Frierson et al. report a mean survival of 4 months with no disease free patients [2]. Other studies report mean survival under 18 months with 5 yr survival in less than 20% cases [4-6]. Local recurrence is common and is the major cause of morbidity and mortality.Unfortunately in our case too patient survived only 4 months after the diagnosis even after receiving cobalt teletherapy.

Conclusion

Sinonasal undifferentiated carcinoma is a very rare tumour with fewer than 100 reported cases. SNUC arises from the Schneiderian epithelium and presents with overlapping clinical and pathologic finding. In limited biopsy material differentiation of the tumour types may be challenging. In our case too diverse histopathological diagnosis was made in the single tumour as TCC (September 2014) and SNUC (April 2015, final diagnosis) when section was taken from fully resected sinonasal mass. The picture was consistent with Inflammatory Polyp (March 2015) on punched biopsy specimen was examined.

So, correct diagnosis necessitates use of adjunct studies like IHC that plays prime role in the diagnosis.

References