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Case Report
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Biphenotypic Sinonasal Sarcoma Successfully Treated with Complete Excision and Facial Reconstruction with Flap: Case Report and Review of the Literature

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Abstract

Biphenotypic sinonasal sarcoma is an extremely rare soft tissue sarcoma that arises in the sinonasal cavity and only 36 cases have been reported. It is a low grade sarcoma of the upper nasal and sinus cavities, specifically the frontal and ethmoid sinuses, predominantly occurring in middle aged females. The present case report describes the occurrence in a middle aged patient and the successful reconstructive facial surgery with the patient doing well 36 months later without recurrence. Biphenotypic Sinonasal Sarcoma successfully treated with complete excision of the tumor and facial reconstruction with flap: Case report and review of the literature.

Introduction

Many different malignancies occur in the head and neck region and each requires different treatment modalities. Biphenotypic Sinonasal Sarcoma (BSNS) has recently been added to this list by the newest WHO classification of tumors of the head and neck [1,2]. Tumors of the head neck region are varied in composition and each require different treatment modalities but sarcomas are extremely rare and surgery the mainstay of treatment.

Case Presentation

A 52 years female presented with 3 months history of right sided recurrent nose bleeds and 1 month history of bulge of right nasal sidewall. On physical exam, patient was noted to have fullness of right nasal ala/sidewall and some excoriation of right anterior septum. Biopsy was performed at that time and histologically demonstrated a malignant spindle cell neoplasm with neural and myogenic differentiation. Immunohistochemistry was positive for S-100, NSE and vimentin. SMA equivocal, desmin, pancytokeratin, synaptophysin, chromogranin, HMB-45, Melan A were negative and these findings were consistent with biphenotypic sinonasal sarcoma. After the diagnosis a CT scan with contrast was then performed showing some mucosal thickening of right anterior septum and inferior turbinate. PET scan showed mildly increased FDG uptake of right anterior nasal cavity consistent with lesion *vs.* post-biopsy changes. Patient was taken to operating room for excision with plan for delayed reconstruction to ensure adequate margins prior to reconstruction. Lesion was excised with defect of 3 cm × 3 cm through and through the nasal ala and sidewall along with 2 cm × 2.5 cm area of anterior septum sparing the contralateral mucosa. Patient was taken back to the operating room 2 weeks later after final path showed concern for 2 close margins. New peripheral margin and septal margin were taken and final pathology confirmed negative margins. Reconstruction was performed after confirmation of negative margins on permanent pathology. Defect was constructed in layers keeping with principles of nasal reconstruction [1]. A cell was used to reconstruct the internal nasal lining and superiorly based mesolabial myocutanous flap including portion of levator labii alaeque nasi used for the soft tissue and cutaneous defect. Levator labii alaeque nasi has previously been described for reconstructions of local defects [2,3]. Patient underwent 54 gy of post-op XRT to primary site starting approximately 1 month post-op. Patient's post-treatment PET scan was negative for any malignancy.

Discussion

There are approximately 68 malignancies that ENT specialists and pathologists encounter in their every day practices of medicine including squamous cell carcinomas, lymphomas, adenocarcinomas, thyroid carcinomas, salivary gland tumors and occasionally metastatic tumors

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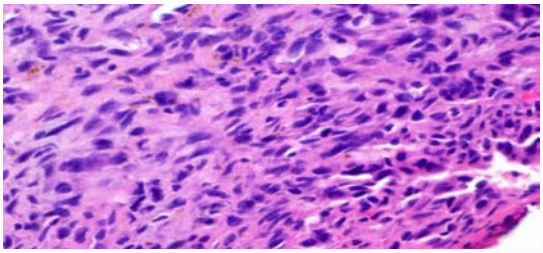

Figure 1: A typical, hyperchromatic spindle shaped cells. H&E stain, 400x.

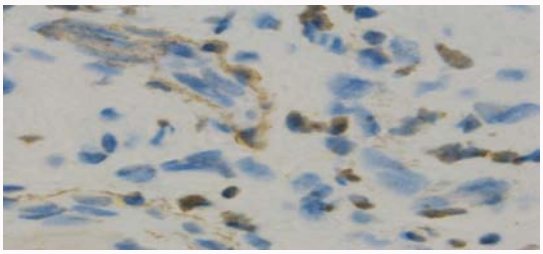

Figure 2: Immunohistochemistry positivity for smooth muscle actin, 400x.

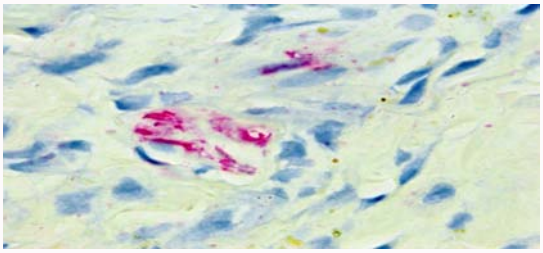

Figure 3: Immunohistochemistry positivity for S100, 400x.

to head and neck; however sarcomas are extremely rare [4,5]. The recent WHO classification of tumors of the head and neck includes several new lesions including NUT Midline Carcinoma (NMC), HPV-related carcinoma with adenoid cystic-like features, *SMARCB1* deficient sinonasal carcinoma, biphenotypic sinonasal sarcoma and renal cell-like adenocarcinoma [6]. Biphenotypic sinonasal sarcoma is an extremely rare soft tissue sarcoma that arises in the sinonasal cavity and only 36 cases have been reported [4]. It is a hypercellular proliferation of uniform spindle shaped cells with minimal atypia and low mitotic activity (Figure 1) [7]. It is named because histologically it demonstrates both myogenic (Figure 2) and neural differentiation (Figure 2 and 3) [8,9] In many cases the myogenic and neural differentiation arises from rearrangements in *PAX3* which allows the cells to differentiate along both cell lines [8-10]. This *PAX3* gene rearrangement has been present in the vast majority, if not all BSNS and is specific for the neoplasm although 4/44 (9%) showed no involvement by either *PAX3* or *MAML3* [4,7,10,11]. It can be associated with *MAML3* and *NCOA1* gene fusions and also *FOXO1* gene fusion [7,9,11]. The myogenic differentiation is demonstrated by positivity to actin and/or calponin and the neural differentiation by positivity to S100 or NSE. The tumor is also positive for Beta-catenin and negative for *SOX10* [12]. The negativity for *SOX10* rules out peripheral nerve sheath tumor [8]. BSNS is negative for cytokeratin in contrast to sinonasal undifferentiated carcinoma and NMC which are positive for cytokeratin and these are high grade neoplasm's presenting in advanced stages and BSNS is not. In head and neck tumors, carcinomas should always be considered and cytokeratin stains performed. Also in the differential are HPV related neoplasms and one of the authors (CON) is extensively involved in researching HPV related neoplasms and testing for HPV should be considered in head and neck neoplasms. Positivity for HPV confirms either HPV related adenoid cystic carcinoma or HPV related squamous cell carcinomas and rules out NMC. The tumor is locally aggressive with recurrence rates up to fifty percent but no deaths from the disease and no metastasis and in the present case after 36 months follow-up the patient is free from recurrence [7,13].

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