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NUT Midline Carcinoma in a Pregnant Woman

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NUT midline carcinoma is a rare, highly aggressive tumor that involves midline structures, particularly in the head, neck and mediastinum. It is characterized by NUT gene translocations on chromosome 15. It typically impacts teenagers or young adults, and has a fulminating course leading to death in less than a year in most cases despite aggressive chemotherapy. Due to its location, this tumor is frequently considered inoperable. We present a case of a sinonasal NUT midline carcinoma with orbital invasion discovered during the workup of sinusitis in a young, pregnant woman. The tumor was managed with definitive excision to negative margins followed by aggressive chemoradiation with no evidence of recurrence for 12 months. We propose that diagnosis of NUT midline carcinoma should prompt recognition of the limitations of current medical care and rapid surgical intervention should be undertaken when possible.

**CASE REPORT**

A 32 year old, 27 week pregnant female presented to her primary care doctor for evaluation of a two month history of worsening nasal congestion, facial pressure, and rhinorrhea refractory to antibiotics. Her symptoms also included right periorbital edema without vision changes. Her medical history was significant for hypothyroidism, but no history of immunosuppression or malignancy. There was no history of prior tobacco use. Given her worsening symptoms despite maximal medical therapy, a CT of the midface (Fig 1) was obtained, demonstrating a large right-sided nasal mass involving the adjacent paranasal sinuses and right orbit. MRI of the orbits, face, and neck (Fig 2) was obtained for soft tissue characterization, and further demonstrated right orbital extension inferiorly and medially, with tumor adjacent to optic nerve. Pre-operative imaging also revealed the presence of an enlarged right level II node.

**TREATMENT**

Biopsy of the mass showed a malignant epithelial neoplasm with sarcoidous features. Immunohistochemistry was carried out, demonstrating the markers of sarcoidous differentiation, p16, and NUT antibody markers. Both histologic features (Fig 3A) and immunohistochemical profile (Fig 3B) confirmed the diagnosis of NUT midline carcinoma.

Given the complexity of the diagnosis in a young, pregnant patient, this case was reviewed extensively at a Multidisciplinary Tumor Board at Jefferson Head and Neck Surgery in Philadelphia, PA. This case was of substantial treatment and history of poor response to traditional chemoradiation in association with the typically fulminant course of NUT midline carcinoma led to a consensus recommendation for aggressive surgical extirpation when at all possible. Right orbital/ethmoidal dissection with temporalis muscle flap and resection of the left maxillary, ethmoid, and sphenoid sinuses was performed. The histology section samples confirmed intraorbital invasion into both inferior oblique and inferior rectus muscles. Extensive sinonasal resection was carried out, with removal of involved skull base structures, including the orbitomeatal plate, fossa ethmoidalis, and underlying dura. Bilateral modified radical neck dissection of levels I-IV was carried out. Given the extent of the resection, harvest of an anterolateral thigh free flap was undertaken for reconstructive purposes. A multidisciplinary team was present in the operating suite for continuous facial monitoring. Both the patient and the fetus remained stable throughout the case and experienced no intraoperative or postoperative complications. All surgical margins were negative on final pathology and neck lymph nodes showed no evidence of local spread. At 36 weeks gestation, birth was induced with uneventful delivery of a healthy infant. Postpartum PET/CT scan showed no evidence of metastatic disease. The patient underwent adjuvant therapy including three cycles of platinum-based chemoradiotherapy and 60Gy of intensity-modulated radiation therapy to the primary site, as well as 54Gy to the right neck over 30 fractions. Surveillance PET/CT scan undertaken since surgery have so far shown no evidence of recurrent or metastatic disease. The patient remains disease-free at 12 months.

**REFERENCES**


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