NUT Midline Carcinoma in a Pregnant Woman

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NUT midline carcinoma in a Pregnant Woman

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ABSTRACT

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 chemoradiotherapy. 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 chemoradiotherapy 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 therapy 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 rhinorhea 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 squamous features. Immunohistochemistry was carried out for the markers of squamous differentiation, p16, and NUT antibody markers. Both histologic features (Fig 3A) and immunohistochemical profile (Fig 3B) confirmed the diagnosis of NUT midline carcinoma.

Fig 3. A. Influenza, high grade squamous carcinomas showing abrupt transition from undifferentiated cells to well differentiated, mature squamous cells. B. Tumor nests, nuclear atypia and brisk activity are present (hematoxylin and eosin, original magnification ×400).

Given the complexity of the diagnosis in a young, pregnant patient, this case was reviewed extensively at a Multidisciplinary Tumor Board. The lack of standard treatment and history of poor response to traditional chemoradiation in association with the typically fulminating course of NUT midline carcinoma led to a consensus recommendation for aggressive surgical resection. Right orbitotomy with cranio-orbital bone flap and trachectomy for airway protection and subsequent resection of the tumor under general anesthesia were carried out. Postoperative histopathological sections 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 orbitofrontal plate, fovea ethmoidalis, and underlying dura. Blalock modified radical neck dissection of levels I-IV was carried out. The extent of the resection, harvest of an anteriorly based thigh free flap was undertaken for reconstructive purposes. A multidisciplinary tumor team was present in the operating suite for continuous fetal 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 chemotherapy 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 scans undertaken since surgery have so far shown no evidence of recurrent or metastatic disease. The patient remains disease-free at 12 months.

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REFERENCES


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