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Case Report

Navigating dual vulnerability: a rare case of esthesioneuroblastoma in pregnancy

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ABSTRACT

Esthesioneuroblastoma (ENB) is a rare malignancy of the sinonasal tract from the olfactory neuroepithelium. Our purpose is to highlight this scarcely described but aggressive tumor presenting in a unique setting where the patient is facing dual vulnerabilities: pregnancy and EBN. We report a case of a 29-year-old female (G2P1L1), 24 weeks pregnant, with a history of progressively increasing left side nasal mass with proptosis, diplopia, nasal discharge, and bleeding from the mass. MRI showed a polypoidal mass in the left nasal cavity with intraorbital and intracranial extensions. The biopsy revealed Hyams grade 2 ENB. Management included craniofacial resection of the lesion and chemoradiation. Caesarean section was performed at 32 weeks, and the patient delivered a baby with a birth weight of 1.463 kg. Early diagnosis and timely intervention, combined with a multidisciplinary approach, for craniofacial resection and chemoradiation, can yield favourable outcomes in this challenging condition.

Keywords: Esthesioneuroblastoma in pregnancy, Olfactory neuroblastoma, Sinonasal tract tumor, Pregnancy

INTRODUCTION

Esthesioneuroblastoma (ENB), or olfactory neuroblastoma (ONB), is a rare neuroectodermal malignant tumor originating from the olfactory neuroepithelium of the sinonasal tract.[1] These cells are particularly dense around the upper part of the nasal cavity, the roof of the nose, and the cribriform plate. Consequently, nasal obstruction is the most common symptom, often accompanied by nasal bleeding, with the tumor tending to invade through the skull base and orbit.[2] Clinical features of locally advanced disease include anosmia, proptosis, pain, or headache. These non-specific presenting symptoms make diagnosis challenging. It has no known sexual predilection and exhibits a bimodal age distribution, with increased occurrence in the second and sixth decades of life.[2] Microscopically, ENBs are composed of neuroblastoma cells and morphologically have a lobular architecture.[3] The Kadish staging system, modified by Morita and colleagues, is commonly used. A cervical lymph node or distant metastases indicate the most advanced stage stage D.[4] Hyams et al proposed a grading system that classifies tumors into four groups based on mitotic activity, nuclear pleomorphism, rosette formation, necrosis, and fibrillary matrix features.[5] Prognosis depends on the extent of disease and tumor grade.[6] We present a case of a pregnant woman with a locally advanced ENB.

CASE REPORT

A 29-year-old, 24-week pregnant G2P1L1 woman presented to our centre with complaints of progressively increasing nasal mass, proptosis of the left eye, diplopia, left facial pain, nasal discharge, and bleeding from the mass (Figure 1A). The patient had a history of spontaneous

conception and a previous uneventful pregnancy with normal vaginal delivery.

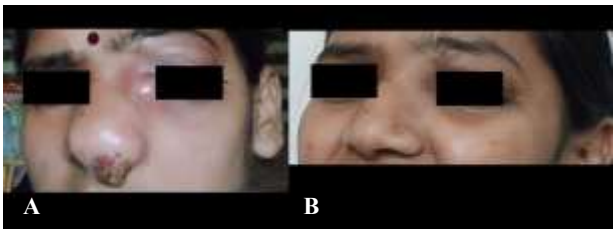

Figure 1: (A) Clinical photograph of the patient at the time of presentation showing a nasal mass protruding through the left nostril, discharge, and left eye proptosis and (B) clinical photograph of the patient after endoscopic craniofacial resection showing a significant reduction in mass size and resolution of proptosis.

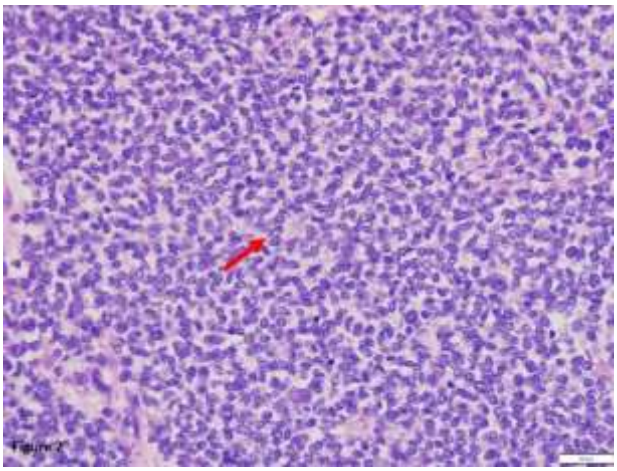

Figure 2: High power magnification of a histopathology slide from the resected mass lesion showing monomorphic and moderately enlarged cells with round to oval nuclei, stippled chromatin, inconspicuous nucleoli, scant cytoplasm, and Homer-Wright pseudo-rosette formation (red arrow).

A biopsy was performed, revealing a cluster of tumor cells surrounding vessels, separated by a fibrocollagenous stroma, with fine chromatin, vacuolated cytoplasm, and a minimal fibrillary matrix in the background (Figure 2). Immunohistochemistry (IHC) showed positivity for synaptophysin. On physical examination, the patient did not have cervical lymphadenopathy.

The cranial and cervical magnetic resonance images (MRI) revealed a large polypoidal mass with lobulated margins in the nasal cavity, predominantly on the left side, with osseous destruction of the ethmoid bony septae, intraorbital and intracranial extensions eroding the left cribriform plate, and infiltration into the left frontal lobe. Left side proptosis and retained secretions in the left sinuses were also noted (Figure 3A). The patient was diagnosed with ENB, Hyams grade II, modified Kadish stage C.[5,6]

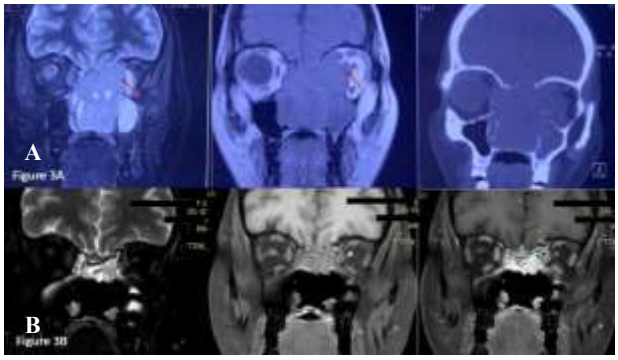

Figure 3: (A) MRI scan before surgery showing a large poypoidal mass with lobulated margins in the nasal cavity with intraorbital and intracranial extension and erosion of the cribriform plate with infiltration into the left frontal lobe (red arrow) and (B) MRI scan after surgery showing a significant reduction in mass size with a 2.7×2.7×2.3 cm residual mass near the cribriform plate (blue arrow).

The available management options, including surgery, external beam radiotherapy (EBRT), and chemotherapy, were evaluated, and pregnancy was planned to be continued with risks like blood loss, preterm labour, hypoxia affecting the fetus due to surgery, and the risk of fetal growth restriction, neurodevelopmental abnormalities due to radiotherapy. Chemotherapy has a relatively low risk to the fetus during the second and third trimester.[7]

The patient was planned for craniofacial resection of the lesion under general anaesthesia, followed by chemoradiation. The patient developed gestational hypertension and intrahepatic cholestasis of pregnancy in the post-operative period. After a non-reassuring non-stress test (NST), the decision to terminate the pregnancy by Cesarean section was made at 33 weeks of gestational age. Patient delivered a baby with a birth weight of 1.463 kg and an APGAR score of 7,9. The baby's further course in the hospital was uneventful.

Despite a significant reduction in mass size on gross examination (Figure 1B), the post-operative MRI showed a residual lesion measuring 2.7×2.7×2.3 cm, for which further EBRT and chemotherapy were planned (Figure 3B).

DISCUSSION

ENB typically presents with nonspecific symptoms, primarily of nasal origin, including obstruction, bleeding, anosmia, and pain. The sporadic occurrence adds to the challenges in diagnosis, along with non-specificity, causing a delayed presentation of most cases of advanced ENB. Histologic differentials for this tumor also include small round cell malignant neoplasms. Immunohistochemistry (IHC) features, including positivity for neuron-specific enolase (NSE) and

synaptophysin, favour the diagnosis of olfactory neuroblastomas.[8] A variety of tumors, including neuroendocrine carcinomas, melanoma, rhabdomyosarcoma, sinonasal undifferentiated carcinoma, lymphoma, and metastatic tumors, are the usual differentials considered during the diagnosis process.[9] Chemoradiotherapy following surgery is regarded as the most effective treatment strategy, based on the limited data available in the literature regarding this condition.[10] The metastasis to cervical lymph nodes indicates a poorer prognosis.

Our case report presents a 29-year-old, 24-week pregnant G2P1L1 woman with Hyams grade II, modified Kadish stage C, T3N0M0 ENB. The locally advanced, painful mass lesion with invasion of the orbit and cranium was planned for surgery followed by chemoradiotherapy.

Surgery remains the mainstay of treatment due to the rapid improvement in compression symptoms and histopathologic examination. Tumors with intracranial and orbital extensions require a multidisciplinary approach. Craniofacial en bloc resection in such cases has been shown to offer better local control and fewer local recurrences as compared to other surgical resections.[11] Kadish stage A tumors can be managed with surgery alone. We proceeded with endoscopic craniofacial surgical resection in our case.

The role of adjuvant radiotherapy (RT) is paramount due to the difficulty in delineating surgical margins and the often locally infiltrating nature of the excised mass. Adjuvant radiotherapy is instrumental in reducing the incidence of local recurrences, especially in patients with Kadish stages B and C.[12] RT delivery is targeted at the tumor bed. At the same time, nodal irradiation is used in cases with lymph node involvement. While the dose of RT varies between 50-60 Gy, higher doses are known to be causative of neural toxicity.[13] The principal risks of RT during pregnancy include fetal malformations, mental disability, and radiation-induced cancer. However, supradiaphragmatic irradiation causes minimal risk to the fetus.[14]

The role of adjuvant chemotherapy is rather significant in locally advanced or metastatic tumors than in early-stage tumors. It is often combined with radiotherapy to decrease the size of the tumor, relieving compression effects and thus aiding in the complete removal of the tumor. The commonly used agents in chemotherapy include cisplatin, etoposide, Adriamycin, vincristine, and cyclophosphamide. The literature lacks ample data about chemotherapy in ENB during pregnancy. Chemotherapy during the second and third trimesters is reported to be safer in terms of risk of foetal malformations as compared to exposure during the first trimester. A study measuring cisplatin placental transfer noted negligible transfer across the placenta at term, suggesting a minimal risk to the fetus with cisplatin use in pregnant patients.[15] The role of pretreatment before each cycle with a dopamine D2 antagonist and 5-hydroxytryptamine (5-HT3) receptor antagonist is vital in the prevention of chemotherapy-induced nausea.

CONCLUSION

Our case attempts to highlight challenges in the management of locally advanced ENB in a pregnant woman. From timely diagnosis with histopathology to proceeding with treatment, keeping in view the safety of both the mother and fetus is crucial in the management of these cases. Endoscopic craniofacial resection is a safe and effective method of removing locally advanced mass lesions with minimal chances of local recurrence in such cases. Radiotherapy in supradiaphragmatic post-surgical resection remnant mass carries minimal fetal risk. Chemotherapy with a cisplatin-based regimen is considered safe in the third trimester as the organogenesis is already complete, and transplacental transfer is also significantly less. It is essential to maintain long-term follow-up for these patients due to the risk of local recurrence.

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