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Esthesioneuroblastoma: A Rare Case Report

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Abstract: ***Esthesioneuroblastoma is a rare, malignant neoplasm of neuroectodermal origin in the roof of nasal fossa with incidence rates of less than 5% of nasal cavity tumors. It shows a variable course from being indolent tumor to highly aggressive neoplasm depending upon various factors e.g, kadish classification, hyam grading of tumor, presence of lymph nodal involvement, metastasis, tumor proliferation index, extent of surgical resection, p53 overexpression and deletion of chromosome 11. The clinical manifestations are non specific and include nasal obstruction, epistaxis, hyposmia, headache and exophthalmos. Surgery is the mainline of treatment followed by adjuvant radiotherapy. Despite treatment recurrence rates are as high as 60 % with mean 5 year survival rates of 33%- 87%. We present a post operative case of 27 year old male patient with left proptosis and esthesioneuroblastoma of KADISH type B.***

Keywords: Esthesioneuroblastoma, malignant neoplasm, craniofacial resection, radiotherapy

1. Introduction

Esthesioneuroblastoma is an extremely rare, malignant neoplasm of neuroectodermal origin that arises from neuroepithelial cells of the olfactory membrane in the roof of nasal fossa. It frequently extends from the upper part of the nasal cavity to the upper part of the septum, the upper nasal conchae, the roof of the nose, and the cribriform plate of the ethmoidal sinus. It represents less than 5% of nasal cavity tumors with intracranial extension seen in 30-40% of cases [1]. It is also known as olfactory neuroblastoma and can show variable course from being indolent tumor to highly aggressive neoplasm.The clinical manifestations are non specific and include nasal obstruction, epistaxis, hyposmia, headache and exophthalmos with most of the patients presenting in locally advanced stage. The diagnosis is made by pathological analysis and immunohistochemical staining. Depending upon stage, treatment ranges from craniofacial resection to minimal invasive approach followed by radiotherapy [2]. We present the case of a 27-year-old male patient, who was diagnosed with esthesioneuroblastoma. He was treated by surgical resection followed by adjuvant chemoradiation.

2. Case Report

27 year old male presented with chief complaint of proptosis of left eye since 1 month. There was no other neurological deficit on examination. CECT findings were suggestive of large homogeneously enhancing lesion involving bilateral nasal cavities and extending into nasopharynx through left choana compressing left frontal lobe and medial rectus causing left eye proptosis. Tumor also infiltrated left maxillary sinus, sphenoidal and ethmoidal sinus. Patient underwent craniofacial resection through bi-coronal approach. Histopathological examination revealed malignant small round cell tumor with immunohistochemistry positive for CD56, synaptophysin and cytokeratin. The features were consistent with olfactory neuroblastoma – Hyams Grade IV. Post op imaging showed large residual tumor of 5 X 6 cm. Adjuvant radiotherapy was given in the dose of 60Gy in 30# for 6 weeks by VMAT along with weekly inj cisplatin in the dose of 30mg/m^2. However, due to high grade of the tumor, the disease showed progression even after completion of radiotherapy. The patient was started on palliative oral chemotherapy based on capecitabine and cyclofosfamide and is undergoing the treatment till date.

3. Discussion

Esthisioneuroblastoma also known as olfactory neuroblastoma arises from neural crest cells of upper nasal cavity [3]. It can occur at any age but bimodal age distribution has been classically described: between 10 and 20 years of age, and between 50 and 60 years with no sex predilection [4,5]. No risk factor has been clearly identified in the literature,however, some studies suggest a possible role of nitrosamines, wood dust and certain genetic abnormalities (3p-); (17q+) [6]. Most common symptoms of olfactory neuroblastoma are nasal obstruction with recurrent epistaxis and in locally advanced stages orbital symptoms like proptosis, diplopia, epiphora. In our case, 27 years old male presented with proptosis with KADISH TYPE B tumor. Lymph nodal involvement is seen in 20% - 25% of cases [7]. Neurological signs are rare. Metastasis occurs mostly to lungs and bones by hematogenous as well as lymphatic route.

High resolution Computed tomography scan and magnetic resonance imaging are used as supportive investigations to accurately delineate the extent of the tumor and infiltration into surrounding structures, erosion of bones, intra orbital/intra cranial extension. Olfactory neuroblastoma does not have a specific radiological feature and appears as a homogenous soft tissue mass with uniform contrast enhancement.

The histopathological appearance exhibits homogenous small cells with uniform round to oval nuclei with rosette formation against a fibrillar background. It is often difficult to differentiate it from malignant melanoma, rhabdomyosarcoma, undifferentiated carcinoma and extramedullary plasmacytoma [8] Immunohistochemistry shows positivity for neuroendocrine markers such as- S-100 protein, neuron specific enolase, chromogranin and synaptophysin. In our case IHC was positive for S-100 and synaptophysin and tumor was Hyams grade IV.

Esthesioneuroblastoma can be staged by various classifications e.g, Biller, Dulgerov, kadish, however Kadish classification is the most well accepted staging classification (Stage A, B and C) [9]. Stage A- Disease confined to the nasal cavity, Stage B- Disease extending beyond nasal cavity to one or more paranasal sinuses and Stage C- Disease extending beyond to orbit, base of skull intracranial cavity, lymphnodes or distant metastases.

The management of esthesioneuroblastoma is controversial, due to scarcity of literature.Treatment is mainly surgical, the gold standard being craniofacial resection followed by radiotherapy. Radiotherapy targets the tumor bed as well as the lymph node area, the dose of radiation can range from 45 to 60 Gy in case of large tumor volume [10]. In our case patient was given adjuvant radiation in the dose of 60Gy/30# by VMAT technique along with weekly inj cisplatin in the dose of 30mg/m^2. Irradiation of the regional lymph nodes is indicated in case of metastatic cervical lymph node involvement or prophylactic in the case of locally advanced tumors. Role of chemotherapy is limited in combination with radiotherapy or as palliative chemotherapy in metastatic/ inoperable/ recurrent tumors.

Despite treatment, ENB remains a malignant tumor with a poor prognosis. ENB is a recurrent and metastatic tumor. The recurrence rate has been estimated at 60% of cases [11].The mean 5-year survival ranged from 33% to 87% [12].

The prognostic factors reported in the literature are age, stage according to the Kadish classification, differentiation grade according to Hyams, the presence of lymph node metastases or distant metastases, the limits of tumor removal, the tumor proliferation index, P53 overexpression and recently the overexpression and more recently the presence of a deletion at chromosome 11 [13].

4. Conclusion

Esthesioneuroblastoma is an extremely rare clinical entity with scarce literature available regarding its management. Till date, surgery is the mainline of treatment followed by radiotherapy. Despite treatment, recurrence rates are high and prognosis remains dismal. Disease course, prognostic factors and therapeutic options needs to be explored.

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