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Incidental Finding Of Esthesioneuroblastoma In The Prevelance Of Mucormycosis

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Abstract

Esthesioneuroblastoma is an uncommon tumor of neural crest origin arising in the nasal cavity. Since 1966, 97 cases have been reported in the world literature[1]. In this report an analysis is presented of the following aspect: age and sex distribution, disease staging, treatment results, interval to recurrence, and survival. During the pandemic of COVID-19 in the timing of second wave in 2021 , mucormycosis cases had started rising , a case of esthesioneuroblastoma was diagnosed with the same findings. Treatment consisted of radiotherapy, surgery, or a combination of radiotherapy and surgery.

Keywords: NIL

Introduction

Esthesioneuroblastoma, is a rare cancer of the nasal cavity. Arising from the upper nasal tract, esthesioneuroblastoma is believed to originate from sensory neuroepithelial cells, also known as neuroectodermal olfactory cells. Due to the location of the tumor and its proximity to the cranial cavity, esthesioneuroblastoma can be highly invasive and challenging to treat. There is no consensus on an appropriate treatment approach of esthesioneuroblastoma because of the rarity of the disease. Most studies reported cranial surgical resection with radiotherapy or chemotherapy to target the tumor while the treatment of mucormycosis consist of Diagnostic Nasal Endoscopy with Functional Endoscopic Sinus Surgery. Hyam's classifications are an important way of determining prognosis[2].

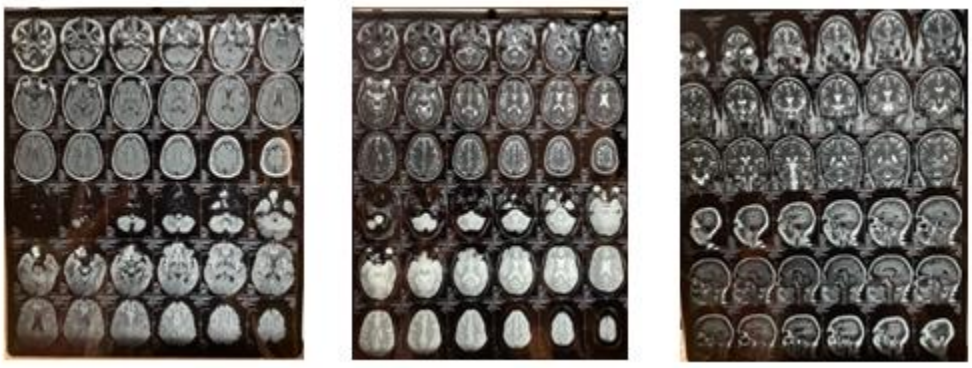
Hyam's histopathological grades for esthesioneuroblastoma

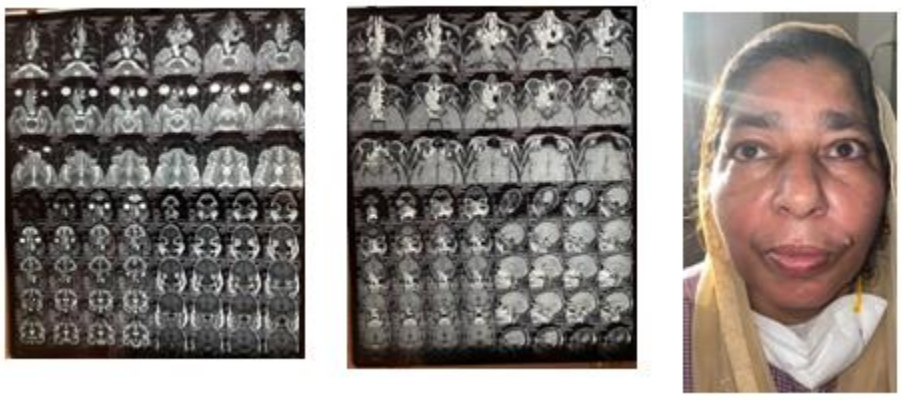
Grade	Lobular architexture preservation	Mitotic index	Nuclear polymorphism	Fibrillary matrix	Rosettes	Necrosis
I	+	None	None	prominent	Homer Wright (HW) rosettes	none

II	+	Low	Moderate	Present	HW rosettes	none
III	+/-	moderate	Prominent	Low	Flexner-Wintersteiner rosettes	rare
IV	+/-	High	Marked	Absent	None	frequent

Case report

A 46y/F with complaints of swelling around right eye and decreased vision in the right eye since 10 days. She gives history of anosmia since 1 month. She also gives history of nasal bleed from right nostril 2 days back, 1 episode, minimal in amount , which stopped spontaneously. She is a known case of Diabetes and Hypertension since 7 years and 4 years respectively, and has been on medications for the same.No history of contact with COVID-19 infected patient. On Anterior Rhinoscopy , deviated septum was noted to the right side. The mucosa was visibally normal and no crusts were seen. On smell test patient could not appreciate any smell. All other Cranial Nerves were normal on examination. Diagnostic nasal endoscopy was done for the patient under local anaesthesia, in which no abnormalities were noted. There was no evidence of mucormycosis was seen. Patient was advised MRI BRAIN [Plain + Contrast].





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MRI Brain(Plain + contrast)- Well defined altered signal intensity soft tissue lesion in the right half of nasal cavity involving right ethmoid and sphenoid sinuses, right superior and middle turbinate's with small extension to the left side, extending into the right orbit through breech involving the right lamina papyracea and surrounding the right retro orbital and intra-canalicular portion of the right optic nerve, involving the adjacent portion of the right superior rectus, medial rectus, superior oblique and levator palpebrae superioris muscles with thinning and focal areas of cortical breech noted involving superior-lateral portions of the body of sphenoid and the right sphenoid sinus and abutting the intra-canalicular portion of the left optic nerve.

Thinning and erosion of the cribriform plate with abnormal meningeal thickening and enhancement involving the basifrontal region.

Above findings could be likely suggestive of esthesio-neuroblastoma (TYPE-C) Lannetti classification Grade IlI)

Diagnosis[edit]

Esthesioneuroblastoma can resemble small blue cell tumors like squamous cell carcinoma, sinonasal undifferentiated carcinoma, extranodal NK/T cell lymphoma, nasal type, rhabdomyosarcoma, Ewing/PNET, mucosal malignant melanoma and neuroendocrine carcinomas (NEC) that occur in the intranasal tract. Compared to other tumors in the region, esthesioneuroblastoma has the best prognosis, with an overall 5 year survival rate of 60-80%.Fewer than 700 cases have been documented in the United States alone. Esthesioneuroblastoma is characterized by neurofibrillary stroma and neurosecretary granules that are not seen concurrently by any other pathologies in the region. On Histological examination tests such as keratin, CK5/6, S-100 protein or NSE can be run to further differentiate esthesioneuroblastoma from other tumors[3].

Staging[edit]

The Kadish classification is used for clinical classification of sinonasal tumors including esthesioneuroblastoma. Subsequent research articles have been published to determine prognosis based on tumor grade[4].

Modified Kadish classification

Stage	Description	5 year survival
A	Tumor confined to nasal cavity	75-91
B	Nasal cavity and paranasal sinuses	68-71
C	Tumor extends beyond nasal cavity and paranasal sinuses, including skull base, orbit or cribiform plate	41-47
D	Tumor metastasizes to cervical lymph nodes and beyond	<40

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Dulguerov classification[5].

Stage	Characteristics
T1	Tumour involving the nasal cavity and/or paranasal sinuses (excluding sphenoid), sparing the most superior ethmoidal cells
T2	Tumour involving the nasal cavity and/or paranasal sinuses (including the sphenoid) with extension to or erosion of the cribriform plate
T3	Tumour extending into the orbit or protruding into the anterior cranial fossa, without dural invasion
T4	Tumour involving the brain
N0	No cervical lymph-node metastasis
N1	Any form of cervical lymph-node metastasis
M0	No metastases
M1	Distant metastasis

Treatment[edit]

The preferred treatment for esthesioneuroblastoma is surgery followed by radiotherapy to prevent recurrence of the tumor. Patient had undergone surgery and started on chemotherapy

Surgical approaches

Several surgical approaches have been described, but post-excision recurrence rates have remained relatively high. Studies suggest better results with a bilateral approach. For cases with cribriform plate involvement, tumors are resected bilaterally using a transfacial and craniotomy approach. In a research study, the craniofacial approach decreased recurrence of esthesioneuroblastoma by 20%[3]. Craniofacial resection can help preserve the optic nerves and brain while removing the cribriform plate, olfactory bulb, dura surrounding the bulb and even the orbital periosteum.

Radiotherapy

Radiotherapy alone is reserved only for small lesions not appropriate for either surgery or chemotherapy. Both photon and proton radiotherapy have been used effectively to treat esthesioneuroblastoma. Proton radiotherapy has recently been shown to be effective in a 10-person study with Kadish C tumors, while delivering less toxicity to the nervous system.

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Chemotherapy

Chemotherapy is used in a multimodality treatment plan generally for more advanced, unresectable or reoccurring
tumors. Cyclophosphamide, vincristine and doxorubi cin have been used as neoadjuvant chemotherapy drugs for grade C esthesioneuroblastoma before surgical resection, producing fair outcomes[6]. Cisplatin and etoposide are often used to treat esthesioneuroblastoma as neoadjuvants or adjuvants with radiotherapy or surgery. Study results are promising. In advanced stage esthesioneuroblastoma in pediatric patients, where surgery is no longer possible, aggressive chemotherapy and radiotherapy has resulted in some tumor control and long term survival.

Prognosis

Esthesioneuroblastoma is a slow developing but malignant tumor with high recurrence rates because of its anatomical position. The tumor composition, location and metastatic characteristics as well as the treatment plan determine prognosis. Common clinical classification systems for esthesioneuroblastoma include the Kadish classification and the Dulguerov classification. Histopathological characteristics on top of Kadish classification can further determine cancer prognosis. In severe, Kadish class C tumors, Haym's grades of pathology are important for prognosis. Patients with low grade Kadish class C tumors have a 10-year survival rate of 86 percent compared to patients with high grade class C tumors who have a survival rate of 28 percent. Surgically treated patients with high grade tumors are more likely to experience leptomeningeal metastases or involvement of the cerebral spinal fluid unlike patients with low grade tumors who usually only see local recurrence[7].

Survival rates for treated esthesioneuroblastoma are best for surgery with radiotherapy (65%), then for radiotherapy and chemotherapy (51%), just surgery (48%), surgery, radiotherapy and chemotherapy (47%) and finally just radiotherapy (37%).From the literature, radiotherapy and surgery seem to boast the best outcome for patients. However, it is important to understand that to some degree, prognosis is related to tumor severity. More progressed, higher grade tumors would result in chemotherapy or radiotherapy as the only treatment. It is no surprise that the prognosis would be worse in these cases.

Incidence

Esthesioneuroblastoma accounts for 2% of all intranasal tumors with an incidence of 0.4 cases per million people. Fewer than 700 cases have been documented in the United States. Fewer than 400 unique cases have been reported globally. Esthesioneuroblastoma can occur at any time, with peak occurrence reported in the second and sixth decades of life

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