Esthesioneuroblastoma presenting with proptosis and cervical lymphadenopathy - An unusual presentation

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Abstract: Introduction - Esthesioneuroblastoma is a rare tumor arising from the olfactory epithelium of the nasal vault which frequently invades the cranial base, cranial vault and orbit. Esthesioneuroblastoma (ENB) presenting simultaneously with proptosis and cervical lymph node metastasis is a very rare presentation. This case report is to discuss the radiation outcome of a patient with Esthesioneuroblastoma, with proptosis and neck metastasis, presented to our Department. Case Report Twenty six year old female presented with proptosis and loss of vision right eye and enlarged cervical lymph nodes for a period of two months whose MRI showed mass in the nasal cavity extending to right maxilla, ethmoid and orbit. HPE showed Esthesioneuroblastoma. Since surgery not possible, the patient was treated with chemo radiotherapy. The tumour completely responded to chemo radiation and proptosis relieved. The cervical lymph nodes also disappeared completely. The purpose of this study is to report the good radiation outcome of this rare nasal tumour with rare presentation.

Keyword: Esthesioneuroblastoma, lymph node metastasis, proptosis, radiotherapy

INTRODUCTION
Proptosis can occur slowly over years, or rapidly over days. Its causes vary geographically, and with the age of the patient. The commonest cause was found to be tumors of both orbital and extra orbital origin from surrounding structures like para nasal sinuses, base of skull and soft tissues of face with intra orbital extension. This case report is to discuss the radiation outcome of a patient with Esthesioneuroblastoma with proptosis and cervical lymphadenopathy, presented to our Department. Esthesioneuroblastoma was first described by Berger and Luc in 1924. These are rare tumors that arise from the olfactory receptors of the nasal mucosa or cribiform plate of ethmoid bone. According to Surveillance, Epidemiology and End Results program 84 cases were reported from 1978 to 1990. There appears to be a slight male predominance.
The age incidence has bimodal age distribution which peaks between 11 to 20 years and 40 to 60 years. These tumours tend to be friable and bleed easily. The most common clinical symptoms are epistaxis and nasal blockage. CT imaging helps to identify the tumour extension to surrounding bony structures whereas MRI will help in assessing tumour spread to surrounding soft tissue areas. A staging system has been proposed by Kadish et al depending upon the extension of the tumour. The optimal treatment of Esthesioneuroblastoma is still debated because of limited number of cases reported. Surgery combined with preoperative or postoperative radiotherapy is widely used in the management of this tumour. For patients with advanced disease where tumour dissemination is more likely, addition of chemotherapy will help in better local control as well as in decreasing distant metastasis. We are reporting an unusual presentation of esthesioneuroblastoma with proptosis and cervical lymph node metastasis and its outcome with chemoradiation.

CASE REPORT
A twenty six year old female presented with proptosis of right eye and enlarged cervical lymph nodes on right side in August 2012. History of occasional bleeding from the eye was also present. The patient also complained of loss of vision of the right eye. There was no history of nasal obstruction or epistaxis. The duration of these symptoms were about two months. On clinical examination, proptosis of the Right eye was present. The right eye globe was fixed in all directions. There was no perception of light in right eye. The left eye was normal. On Anterior rhinoscopy there was a polypoidal mass in the right nasal cavity. Examination of the neck revealed right level 1b, II, III and V cervical lymphadenopathy.

Computerized tomography scan of nose, para nasal sinuses and orbit with contrast showed a heterogeneous lesion involving nasal cavity, bilateral ethmoid sinuses with erosion of the roof of nasal cavity and right medial wall of the orbit. Magnetic Resonance Imaging scan showed heterogeneous lesion involving nasal cavity, right maxillary sinus, bilateral ethmoid sinuses with erosion of roof of nasal cavity, right medial wall of orbit, nasal septum extending into frontal extra axial space. Hence the lesion has been staged as Kadish stage C extension. A biopsy has been taken from the nasal growth and sent for histopathological examination. Histopathological examination revealed small round cell tumor with rosettes suggestive of esthesioneuroblastoma. Fine Needle Aspiration Cytology of submandibular node shows metastatic deposits. The surgical opinion has been sought. Since the tumor is locally advanced with intracranial extension radical excision of the tumor with adequate clearance along with cervical lymph node dissection is highly morbid. Hence the patient was treated with concurrent chemo radiation. Radiotherapy dose of 66 Gy was delivered to tumor and nodes using TELECOBALT machine. The fractionation used was 200 cGy per fraction, five fractions per week for 6.3 weeks. The patient was also given three cycles of concurrent chemotherapy using Cisplatin, Etoposide and Vincristine. The radiation portals to treat primary tumor included one anterior and one right lateral field with wedges. The right side whole neck was included in neck portal. One month after follow up the proptosis completely disappeared. There were no palpable neck nodes on clinical examination.
proptosis - Rt eye

DISCUSSION
Esthesioneuroblastoma is a tumor of neural crest origin arising from the olfactory epithelium in the cribriform plate or nasal cavity. Synonyms include olfactory esthesioneuroma, neuroesthesioma, olfactory neurocytoma, and olfactory esthesioneuroblastoma. There is a bimodal age distribution with peaks at two age groups between 11-20 years and second in 51-60 year olds. On contrary the patient reported in our case is 26 years old. Esthesioneuroblastoma is a friable tumour which bleeds easily. Hence the most common presenting symptoms are nasal obstruction and epistaxis. On contrary our case presented with proptosis and loss of vision of the right eye. There was no history of nasal obstruction or epistaxis. This rare neoplasm is locally aggressive and can metastasize by lymphatic and hematogenous routes. A metastatic rate of 10-30% is reported with the most common site being the cervical lymph nodes similar to our case. According to Kadish staging system proposed for Esthesioneuroblastoma, stage A is disease confined to nasal cavity. Stage B is disease confined to nasal cavity and one or more para nasal sinuses. Stage C involves disease involving orbit, base of the skull, intra cranial extension, cervical

post RT proptosis relieved post RT - neck

Elkon and colleagues at diagnosis is 30% stage A, 42% stage B, and 28% stage C. Of the 22 patients who had stage C disease, 9 had orbital involvement, 6 had invasion of the cribriform plate or cranial fossa, 11 had palpable neck nodes, and only 1 had distant metastasis. Because of orbital involvement, intra cranial extension
Histopathologically, Esthesioneuroblastoma contains epithelial components serving as supporting stroma and neural components that correspond to olfactory cells. Rosettes are the main feature which contains a row of cells arranged around a central area. Also, there will be fibrils which fill the central space of the rosette. These tumours should be differentiated from undifferentiated sinonasal carcinomas which lack rosettes and intracellular fibrils. Hyams proposed a staging system for Esthesioneuroblastoma which includes 4 grades. Grade I tumours have excellent prognosis whereas grade IV tumours are invariably fatal. The Kadish clinical staging and Hyams histological grading are the two most important prognostic factors. In literature, the 5-year recurrence free survival is reported to be between 52% and 90%. Because Esthesioneuroblastoma is rare, there are no standard guidelines for treating this tumour. Various studies have shown that Radiation or Surgery alone is satisfactory for Kadish stage A patients. But with Radiation alone the local control was 58% for stage B and 18.9% for stage C patients at a median follow up of 56.1 months. For locally advanced disease various studies have shown that aggressive multimodality treatment in the form of craniofacial resection followed by adjuvant Radiation provided the best treatment outcome. Overall survival was 55% and 46% at 5 and 10 years of follow-up, respectively. The role of preoperative and concurrent chemotherapy and autologous bone marrow transplant is similarly unclear. For advanced lesions, in which disseminated disease is likely, chemotherapy may improve tumour control and decrease the incidence of distant metastases. A combination of thiotepa, cyclophosphamide, doxorubicin, vincristine, nitrogen mustard, and actinomycin-D has been used in various studies.

**CONCLUSION**

Esthesioneuroblastoma is a very uncommon malignant tumor arising from olfactory epithelium that may have a long natural history characterized by frequent local or regional recurrences. It may have rare presentations including proptosis only or proptosis along with nasal symptoms. Radical craniofacial resections by a multidisciplinary surgical team combined with adjuvant radiotherapy with 50-60 Gy is probably the most usual treatment of Esthesioneuroblastoma. If surgery is not possible, concurrent chemo radiation plays a significant role in tumor control in ENB. Esthesioneuroblastoma is almost a chronic disease, requires very long follow-up, and is one of the few paranasal sinus cancers where surgical salvage at the primary site is possible. We report a case of Esthesioneuroblastoma presented with proptosis and cervical lymphadenopathy and responded well to chemo radiotherapy which improved the quality of life significantly.
REFERENCES


