CASE REPORT

RARE PRESENTATION OF OLFACTORY NEUROBLASTOMA: CASE REPORT

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ABSTRACT: Olfactory Neuroblastoma (Esthesioneuroblastoma) is a rare neuroectodermal tumour. Presentation includes nasal obstruction, recurrent epistaxis, Hyposmia, anosmia, rhinorrhoea. Treatment includes primary excision and/or radiotherapy, chemotherapy. Here we report an unusual case with presentation of mass in nasopharynx with no specific symptoms and incidental cervical nodes.

KEYWORDS: esthesioneuroblastoma, neuroectodermal tumor, Immunohistochemistry, hyposmia.

CASE REPORT: A 35 yr old woman presented to ENT outpatient clinic with multiple neck swellings bilaterally. Minimal discomfort in nasal breathing reported. No other associated symptoms either in neck or nose. Examination revealed multiple non tender firm cervical nodes in level II and III, biggest about 2x2cms. Thorough examination with nasoendoscopy revealed a mucosa covered swelling in the nasopharynx arising from the roof going downwards. MRI^{Fig.2} showed mass confined to nasopharynx, anterosuperiorly extending till choanae. Initial FNAC and later excision biopsy of the neck nodes reported as non specific inflammatory changes. Biopsy from the mass in nasopharynx revealed rosettes^{Fig.1} in the field, reported as possible olfactory neuroblastoma. Immunohistochemistry^{Fig.1} confirmed the diagnosis with the pattern consistent with olfactory neuroblastoma. After clinicopathological review, (owing to non specific nodes) it was decided to send the patient for chemotherapy for which patient responded well with regression of nasopharyngeal mass. Long term follow-up is planned as part of further management.

DISCUSSION: Esthesioneuroblastoma (olfactory neuroblastoma) is a rare neuroectodermal tumour,¹ which is challenging for multidisciplinary management, as so far there is no known accepted standard therapy.² It is thought to arise from the olfactory epithelium.

They are highly malignant and metastasize widely. Seen with bimodal peaks in 2 and 6th decades¹. Presentation can be with nasal obstruction, headaches, sinus problems, facial pain, facial swelling, hyposmia, anosmia, diplopia, haemorrhage or neurological deterioration etc. Examination reveals a mass either in nasal cavity, skull base extending to paranasal sinuses or intracranially. It can also metastasise to cervical lymph nodes.

Microscopically, the tumour is composed of small round cells (slightly larger than lymphocytes) and neurofibrillar matrix, often forming rosettes. Electron microscopy if available shows neural processes and membrane bound secretory granules.

Immunohistochemistry³ shows neuron specific enolase, synaptophysin, S-100 protein positive sustentacular cells, keratin negativity, neuroendocrine marker positivity.⁴ Most of them show 11; 22 translocation and some reveal trisomy 8.

Prognosis is variable. Other differential diagnoses with similar histological appearances but different immunohistochemistry features include Rhabdomyosarcoma, Malignant lymphoma.⁵

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Olfactory neuroblastomas (or esthesioneuroblastomas) are most frequently staged using a system proposed by Kadish et al⁶ in 1976.

- group A : tumour is limited to the nasal cavity
- group B : tumour is limited to the nasal cavity and paranasal sinuses
- group C : tumour extends beyond the nasal cavity and paranasal sinuses
 - base of skull
 - intracranial compartment
 - orbit
 - distant metastatic disease

An additional group D has been proposed by Chao et al⁷ in 2001.

- group D : cervical nodal metastases

Treatment modalities are variable as is the condition. It might include surgery, or chemotherapy (with cisplatin,^{8,9} etoposide⁹ or cyclophosphamide, vincristine with or without Doxorubicin) or radiotherapy or multimodal approach² which is used in majority of cases. Outcome depends on the stage. Recurrence is common. Palliative management done in cases of advanced stages and recurrence.

Here we present an unusual presentation of a case with histologically negative cervical nodes and nasal obstruction.

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