Esthesioneuroblastoma

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Esthesioneuroblastoma is a rare, locally aggressive neoplasm of the sinonasal area, arising from olfactory neuroepithelium and its clinical course is characterized by indolent growth, persistent local recurrence and occasional distant metastases despite attempts at cure. The tumor has no known etiologic cause and the pathologic diagnosis is often difficult because of the wide range of patients' age and histologies. A case of esthesioneuroblastoma is presented both in view of its rarity and to stress in considering it in the differential diagnosis of nasal mass.

CASE REPORT

A fortyfive year old business man presented with

history of left sided nasal obstruction, epistaxis, anosmia, headache and dimness of vision of five months duration. A previous history of nasal obstruction and epistaxis eight years back for which he underwent excision of a mass lesion from his left nostril, and no histopathology was performed. On examination he was anemic, and in his left nostril, a red proliferative mass was seen arising from the roof, which was firm and friable. He had no vision in both his eyes and also had a right lateral rectus palsy. No cervical

Fig.1

*Photo micrograph of Esthesioneuroblastoma
Neoplasm composed of groups of small round cells with dense nuclei, scanty cytoplasm and dense fibrous tissue stroma with rosette and pseudorosette formation (H&E staining)
Magnification 400.*

Fig.2

*CT Scan of brain and paranasal sinuses – contrast study
Shows an expansile soft tissue mass involving the nasal cavities, all paranasal sinuses on both sides, with extension to frontal lobe and suprasellar region.*

lymph nodes were palpable.

A punch biopsy was taken and the histopathology revealed Esthesioneuroblastoma. It is Neoplasm composed of groups of small round cells with dense nuclei, scanty cytoplasm and dense fibrous tissue stroma with rosette and Pseudorosette formation (see fig.1).

A CT Scan of his brain and paranasal sinuss was taken to assess the extent of the lesion for considering surgery. It showed expansile soft tissue mass involving the nasal cavities, all paranasal sinuses on both sides, with extension to orbital apices bilaterally and extension to frontal lobe. There is also evidence of suprasellar extension (see fig.2).

Due to the extensive regional and intracranial spread of the tumour, we planned a craniofacial resection and removal of the tumour under general anaesthesia. But due to the added risk of the procedure, the patient was unwilling for the surgery. Hence he was referred for radiation treatment he was given a total dose of sixty Gy over a period of six weeks and he is being followed up.

DISCUSSION

Esthesioneuroblastoma is a rare malignant tumour of nasal cavity. It was first reported by Berger in 1924. Since then, more than 200 cases of Esthesioneuroblastoma have been added to the world literature (1990).

The overall incidence is 0.25% of all intranasal tumours. It has a sex predirection in males and it occurs in all age groups with bimodal peak in second and sixth decades.

This is also called as esthesioneuroepithelioma, esthesioneurocytoma, olfactory esthesioneuroma, intranasal neuroblastoma, neuro olfactory tumour and olfactory placode tumour. Ogura and Schenek have referred to this tumour as the "great impostor", since it has often been mistaken for lymphoma, melanoma, undifferentiated carcinoma and Sarcoma.

The tumour is believed to have a neuroectodermal origin. It arises from the sustentacular cells of olfactory epithelium in the superior third of nasal septum, the cribriform plate and superior turbinate regions. Kadish developed a classification system based upon the clinical extent of disease at presentation. Stage A, disease confined to the nasal cavity: Stage B, disease confined to nasal cavity and paranasal sinuses : an Stage C, disease spread beyond nasal cavity an paranasal sinuses. Hyams attempted to correlate survival with a four part histopathologic grading system based largely on the level of differentiation.

Clinically these tumours are usually unilateral, broad-based, red, fleshy and friable masses. They vary in consistency from soft to hard and are slow growing in nature. The tremendous age range of patients at presentation coupled with the varied histologic picture results in a wide differentail diagnosis.

The tumour can locally extends to paranasal sinuses, orbit, palate, anterior or middle cranial fossae. There's twenty percentage chance of metastasis to cervical lymphnode, lung, thyroid and bone. Apart from the standard histological diagnosis, immunohistochemical study can be useful using the stains cytokeratins, neuron specific enolase, synaptophysin, chromogranin, S-100 protein etc. Recurrence can be monitored by urinary assay of Dopamine and VMA.

The mainstay of treatment of this tumour is surgery followed by radiotherapy. According to the extension of the lesion, various surgical procedures have been advocated. Radiotherapy alone is advocated for residual or recurrent tumours, highgrade tumours, and low grade tumours when margins are close. Recent advances in treatment includes combined chemotherapy and proton radiation and is mainly advocated for high grade and inoperable tumours. This protocol has an acceptable complication rate and conveys less morbidity than cranio facial resection and conventional radiotherapy. High dose chemotherapy followed by autologous bone marrow transplantation

should be considered as a salvage regimen for patients who fail conventional therapy.

In conclusion the otolaryngologist should possess a high index of suspicion for this rare but

devastating malignancy. This constant vigilance, coupled with the opportunity for rapid clarification of diagnosis through the use of immunohistochemical techniques may enhance the early and accurate diagnosis of this "Great

References

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CONFERENCE NEWS

Workshop on Functional Endoscopic Sinus Surgery

Department of Ear, Nose & Throat, Batra Hospital & Medical Research Center, New Delhi & Dept. of Anatomy, Santosh Medical College Ghaziabad, Dec 5–7, 1997.

A Report

The first workshop on Endoscopic sinus surgery was organised by the Department of Ear, Nose & Throat, Batra Hospital & MRC. Prof. S.K. Kacker was the workshop Director and Dr. Ajit Mann Singh was the Organising Secretary. The operating Faculty was Prof. S.K. Kacker & Dr. Ajit Mann Singh from BH&MRC, and Dr. M.V. Kirtane & Dr. Prabodh Karnik from Mumbai as external Faculty. Dr. Sharat Latta who joined the Department as Consultant only later, functioned De-Facto as Joint Organising Secretary. This workshop was organised under the auspices of A.O.I. Delhi State Branch. Over 40 delegates from all over North India participated in the workshop, 20 of whom went for Cadaver dissection.