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ESTHESIONEUROBLASTOMA - A CASE REPORT

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SUMMARY. ***Esthesioneuroblastoma is an uncommon tumour of the nasal cavity arising from olfactory epithelium. Ever since Berger described the first case of esthesioneuroblastoma in 1924, considerable progress has been made in the diagnosis and management of these tumours. A case report of esthesioneuroblastoma has been presented and relevant literature is reviewed.***

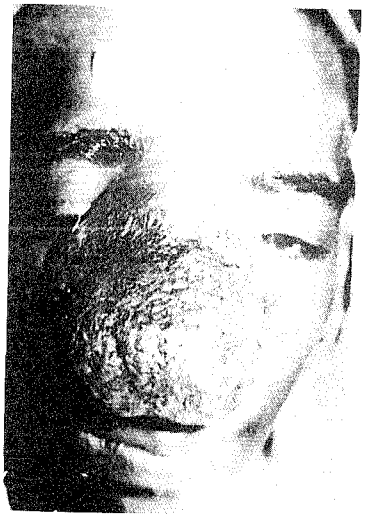

Fig. 1 Pre Operative View

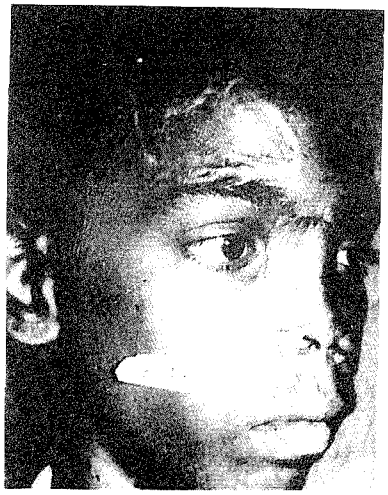

Fig. 2 Post Operative View

CASE REPORT

A 17 year old boy presented with history of swelling arising from left nostril of 2 months duration. The swelling measured 7x5 cm and was found to be involving the tip, dorsum and root of the nose, extending into both the nostrils and was firm and mobile. There was no involvement of regional lymph nodes. Radiological examination revealed soft tissue shadow with erosion of nasal bones. A clinical diagnosis of rhinoscleroma was made, and confirmed by histopathological examination. He was treated by ENT surgeons with systemic antibiotics and local application of acriflavine. The swelling continued to increase in size obstructing both the airways demanding intervention. Excision of the tumour involved, resecting the whole of the dorsum of the nose, four-fifths of alae on both sides, entire collumella, threefourths of the lateral walls and anterior one third of septum. This necessitated total reconstruction of nose. The scalping flap was used for cover and lining was provided with split thickness skin graft. Initially support for the lining was provided with dental stent compound and this was removed after two weeks. At three weeks, the pedicle was cut and the redundant flap returned. Histopathology of the entire excised specimen made us review the diagnosis. After discussion with the pathologist and review of the earlier slides, a diagnosis of esthesioneuroblastoma was made.

Local recurrence was observed at the margin of right alae after 4 weeks. Bone secondaries were also observed 3 months later. Both these problems were managed with radiotherapy, amounting to a total dose of 4,500 rads over a

period of four and a half weeks, in divided doses. At three years he was free of recurrence.

DISCUSSION

Esthesioneuroblastoma or olfactory neuroblastoma is of neural crest origin arising from olfactory epithelium in the nasal cavity. It usually presents high in the nasal cavity in close proximity to the cribriform plate. Most common symptoms are epistaxis and unilateral nasal obstruction. It may present at any age but shows a broad peak of incidence between the second and fourth decades of life.

The overall histological picture closely resembles that of adrenal or sympathetic ganglioneuroblastoma. It is characterised by the presence of round or oval cells of the size of lymphocytes in a neurofibrillary matrix with pseudorosette formation. Perivascular palisading of cells and interstitial calcification are occasionally observed. Electron microscopy and immunohistology have been further suggested for confirmation of diagnosis.

In the present case, the diagnosis was missed during the initial histopathological examination owing to its rarity. Though the tumour was extensive, there was no local extension into paranasal sinuses, orbit or skull bones. Surgical excision was relatively simple, but local recurrence was observed within one month. Radiotherapy has given a very satisfactory result for local recurrence and skull metastases. A 3-year follow up revealed a complete disease free boy except for the problem of atrophic rhinitis. Primary reconstruction with the scalping flap with immediate skin lining has given an aesthetically satisfactory nose.

Clinical staging suggested by Kadish[1] is widely followed. Stage A - involvement of nasal cavity only, Stage B - involvement of nasal cavity and one or more paranasal sinuses, Stage C - extension beyond nasal cavity and sinuses. The clinico-radiological features of the case presented belongs to Kadish Stage C.

Wide excision followed by postoperative radiotherapy is the method of choice for all the resectable lesions.[2] Local irradiation and palliative chemotherapy has been suggested for locally unresectable tumours and for distant metastasis.

Esthesioneuroblastoma is often misdiagnosed clinically and histologically. Only strong suspicion on the part of the clinician and the histopathologist can ensure early diagnosis of this rare tumour. Though local recurrences are common, prognosis is good if the surgical extirpation is adequate and postoperative radiotherapy is given.

References

1. **Kadish S, Goodman M, Wang C C.** Olfactory Neuroblastoma, A Clinical Analysis of 17 Cases. Cancer 1976; 37:1571-1576.
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