Olfactory neuroblastoma

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Abstract

Olfactory Neuroblastoma is a rare, malignant tumour, arising most probably from the olfactory mucosa. It has a strong tendency to invade the surrounding sinuses, the orbits and the cranial fossa and brain. Metastases are frequent.

In spite of its relative radiosensitivity no cure was achieved by irradiation alone. The treatment of choice consists of radical excision combined with irradiation. However, even after clinically successful results of treatment, prognosis is guarded and follow-up imperative.

A case of radioresistant tumour in a 14-year-old boy is presented. The patient is well three years after radical surgery.

The first description of tumours, originating in the nasal cavity and apparently related to the olfactory mucosa, is attributed to Berger and Luc (1924). The reported tumour had a prominent rosette pattern and resembled tumours of the retina. Two years later Berger and Coutard (1926) described another intranasal tumour that formed neurofibrils but not rosettes. They considered it a more mature variant of the previously described type. On that histological basis they assumed that these tumours arose from sensory cells of the olfactory mucosa. Since then increasing familiarity with the clinical and histological features of the olfactory neuroblastoma has been gained, but its histogenesis has not been finally established. The prevalent opinion is that the tumour is of ectodermal origin and arises from olfactory mucosa.

Other possible sources of the tumour considered were: the olfactory placode, Jacobson's vomeronasal organ and the sphenopalatine ganglion.

The olfactory placode, an embryonic progenitor of the olfactory membrane was found unlikely to be the origin of the tumour, as it does not persist in post-embryonic life (Mendeloff, 1957). Jacobson's organ disappears, too, during late fetal development, although it may persist into adult life (Schall and Lineback, 1951). The sphenopalatine ganglion was rejected, too, because its location in the sphenopalatine fossa is inconsistent with the usual site of appearance of the tumour. There is only one report of the tumour located in the nasopharynx (King, 1959).

Case report

A 14-year-old boy was referred to our department after repeated profuse bleeding from the nose for the last 7 months, which gradually resulted in hemorrhagic anemia, vertigo and weakness.

On inspection a broadening of the nasal bridge was striking. There was marked fullness and tenderness of the left cheek and a protrusion of the (L)

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eyeball. In the left nasal fossa a big, firm, whitish tumour was found, pushing the whole nasal septum far to the right, thus resulting in obstruction of both nasal cavities. The nasopharynx was free. In the left supraclavicular area a few mobile, enlarged lymph nodes were palpated, Pulse rate was 120/min., hemoglobin 6·0 gm. per cent, erythrocytes hypochromic, erythrocyte sedimentation rate 40/80 Westergreen. The patient was given a blood transfusion and iron therapy was started.

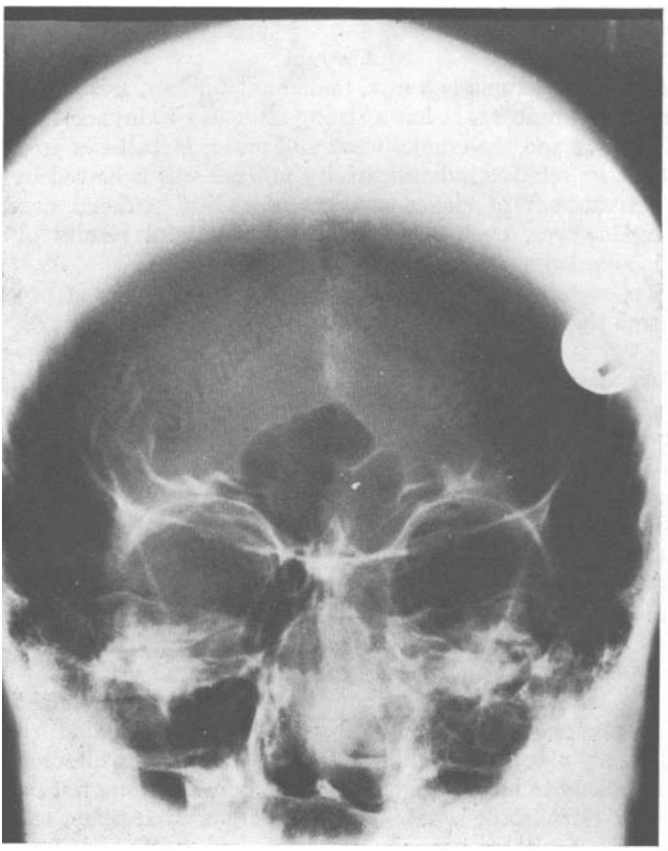


FIG. 1.
X-ray of nasal sinuses, showing a mass in the nose, clouding of ethmoidal and frontal sinuses.

X-ray examination revealed a dense opacity of the left maxillary and ethmoidal sinuses, clouding of the (L) frontal sinus, and destruction of the bony nasal septum and the lower orbital wall (Figs. 1 and 2). Chest and base of skull X-rays were normal. The patient's general condition improved rapidly, after the hemoglobin rose to a level of 13–13·7 gm. per cent. Carotid arteriography showed the internal maxillary artery to be the main feeding vessel of the tumour (Figs. 3 and 4). Biopsy material was obtained from the mass in the nose, from the maxillary sinus by a Caldwell-Luc approach and from the

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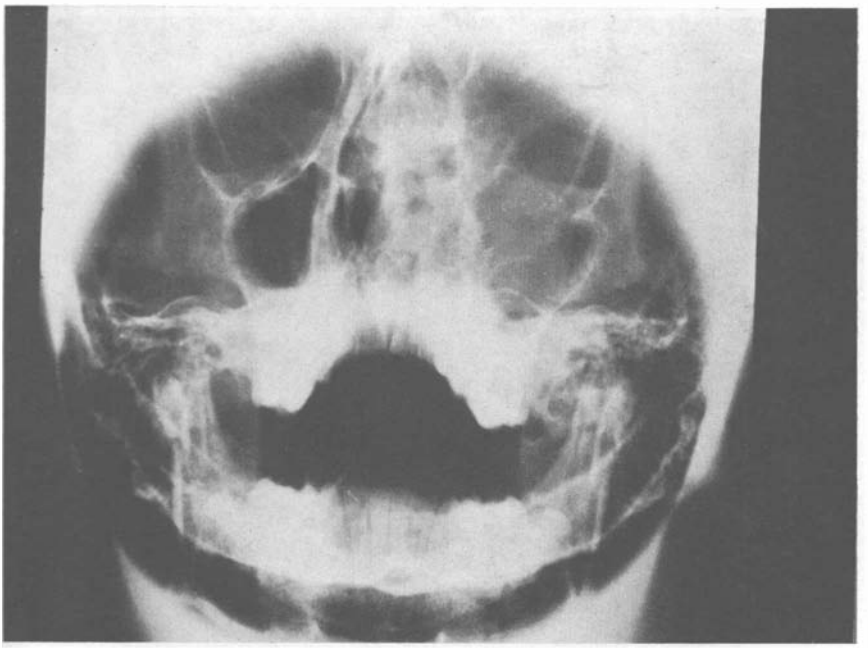


FIG. 2.
Water's view showing tumour in the nose and maxillary-sinus, nasal septum displaced to the right side, destruction of the orbital wall.

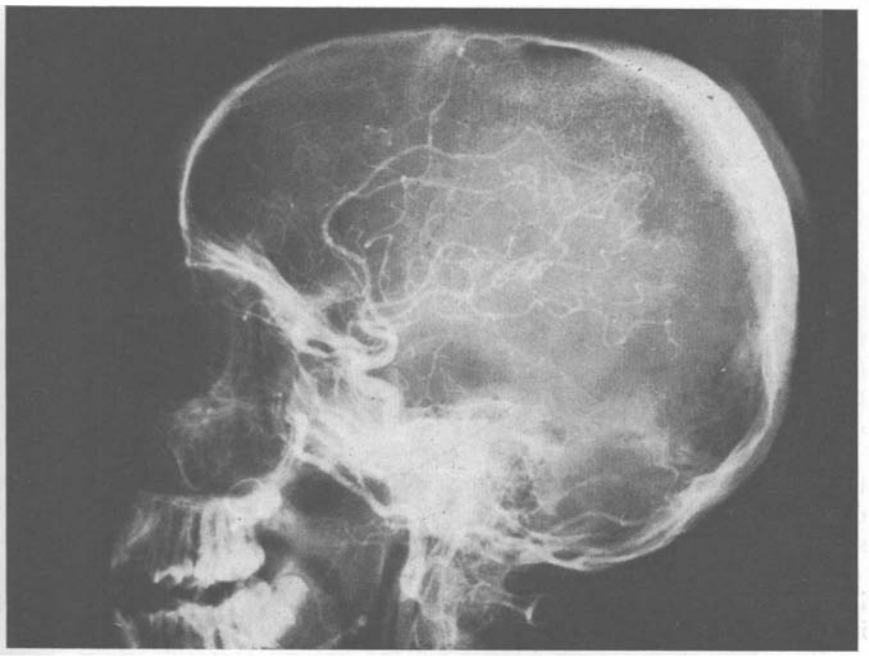


FIG. 3.
Carotid angiogram showing internal maxillary artery as a feeding vessel to the tumour.

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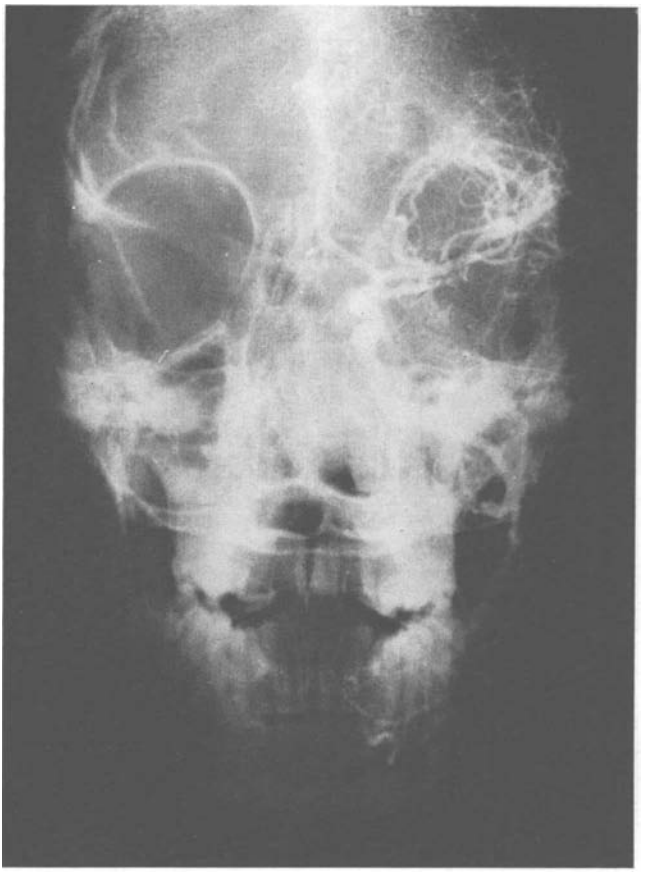


FIG. 4.
Carotid angiogram, postero-anterior view.

frontal sinus by trephination. The biggest of the supraclavicular lymph nodes was also removed for histological examination. Specimens from the nose and nasal sinuses showed the histological picture of an olfactory neuroblastoma (Fig. 5), relatively uniform, small, round or oval neuroblasts, associated with bundles of neurofibrils (Fig. 6). A few rosettes were found (Fig. 7). The lymph node was negative.

Pre-operative irradiation was started with the hope of shrinkage of the tumour in order to make the radical excision less extensive. Total telecobalt dose was 6000 rad in two fields, to the nose and the sinuses, over a period of six weeks.

In spite of this large dose of irradiation there was neither visible change in the macroscopic appearance of the tumour in the nose, nor of its extensions in the X-ray pictures, within the two months following the irradiation.

Operation started with ligation of the external carotid artery. Lateral rhinotomy was performed, the incision included the upper lip. Partial removal

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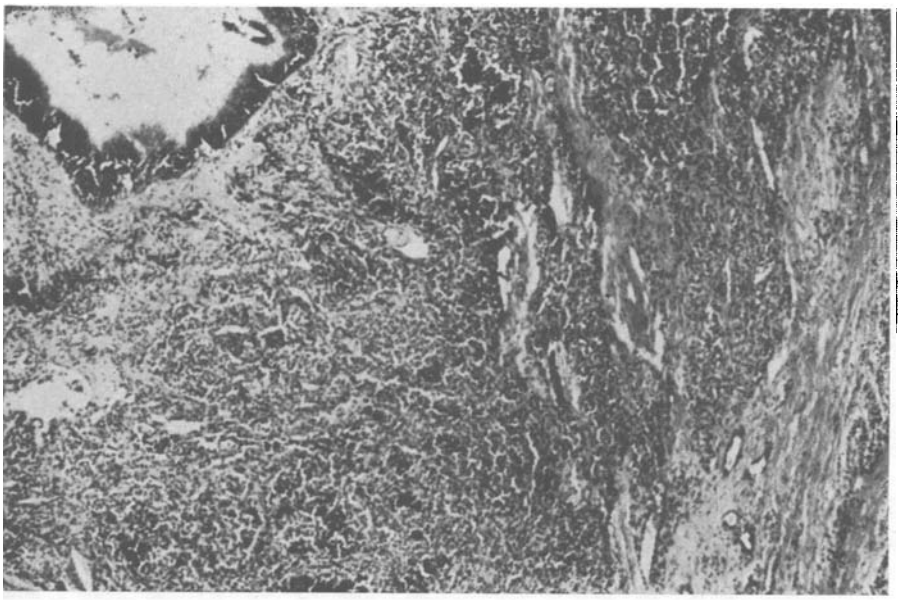


FIG. 5.
Dense tumourous mass growing underneath upper respiratory type epithelial lining.

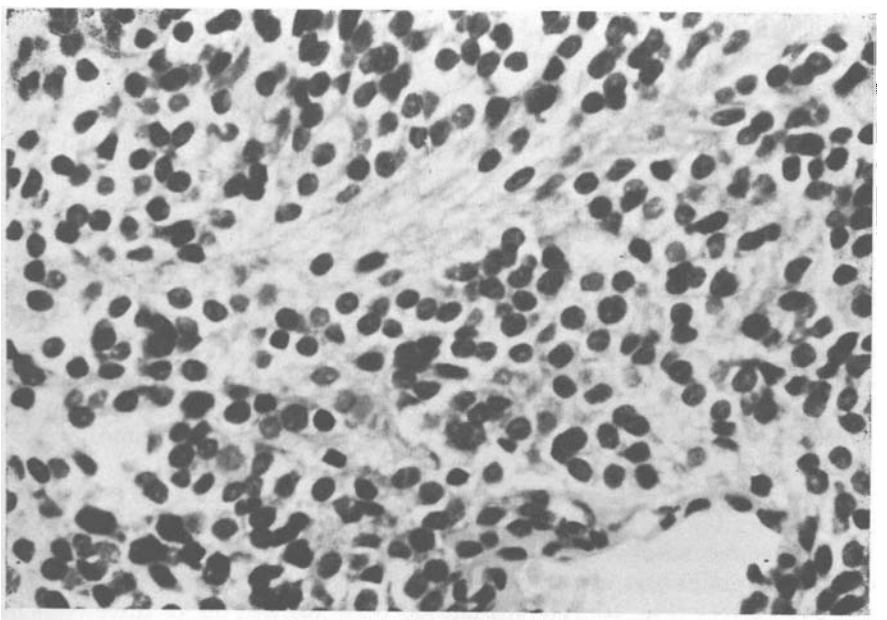


FIG. 6.
The tumour is composed of relatively uniform, small, round or oval neuroblasts associated with bundles of fine neural fibrils.

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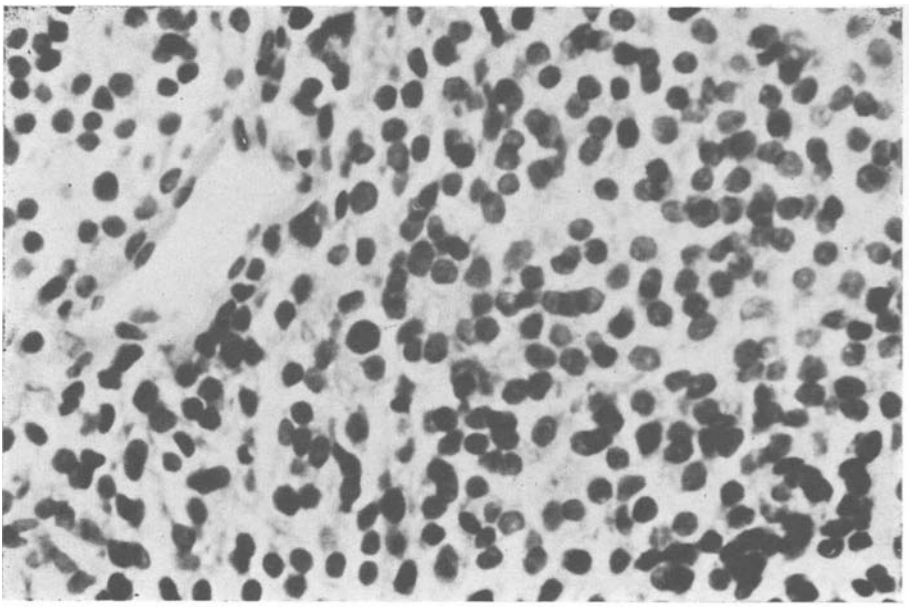


FIG. 7.
One of the rare rosette formations found in this type of tumour is seen in the centre of the field.

of the maxillary frontal process contributed to exposure of the tumour. It extended from the lower medial third of the frontal sinus, anterior ethmoidal cells and cribriform plate downwards and anteriorly toward the nasal vestible hanging a few millimetres above the nasal floor (Figs. 8 and 9). Laterally the tumour extended to about 5 mm. from the lateral wall of the maxillary sinus, destroying its medial wall. The tumour was in contact with the medial third of the orbital floor, which was destroyed, as well as a part of the nasal bone and lamina papyracea. The turbinates of the involved side were not found. The vomer and ethmoidal perpendicular plate were partially destroyed. The tumour was encapsulated and could be easily peeled out from the nasal cavity and the sinuses. The posterior ethmoidal cells and sphenoidal sinus were not invaded. The cribriform plate was inspected in its whole course and seemed undamaged.

The destruction of the bony structures was obviously due to pressure, and signs of infiltration were not noted. A number of biopsies taken from the cribriform plate mucosa and the walls of the invaded sinuses were found negative. No infiltration was found in the removed pieces of bone.

After complete removal of the tumour the septal cartilage was pushed into its normal medial position, the nose packed with Whitehead's varnish tampons, and the wound sutured. The postoperative course was uneventful and the patient was discharged three weeks after the operation. The cosmetical results were satisfactory (Fig. 10). The patient has been followed up for three years by periodic examinations. No local recurrence or metastases have appeared so far. Surprisingly, there is also very little crusting in the post-operative cavity.

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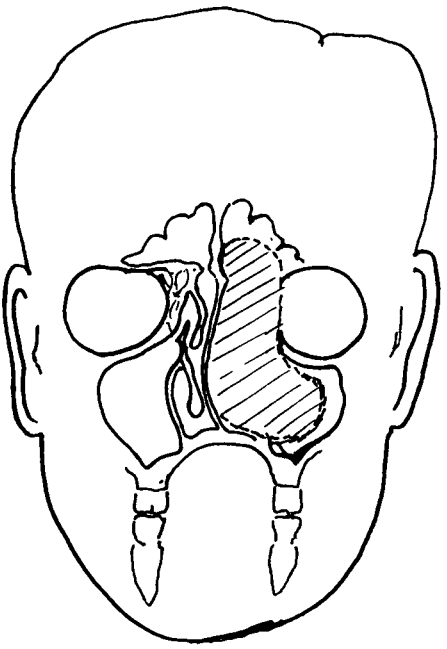


FIG. 8.
Schema to illustrate the operative findings in the coronal plane. Tumour outline dashed.

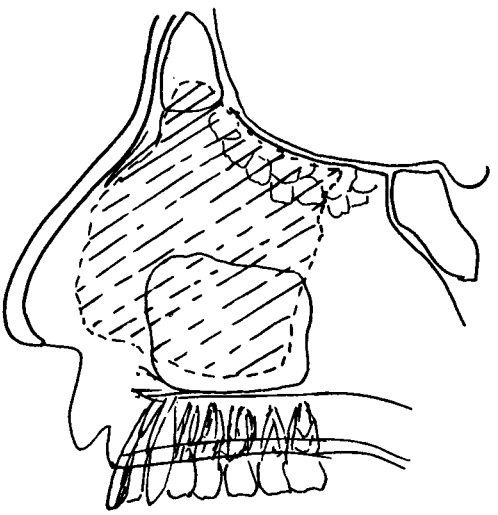


FIG. 9.
Schema showing anteroposterior dimensions of the tumour (dashed area).

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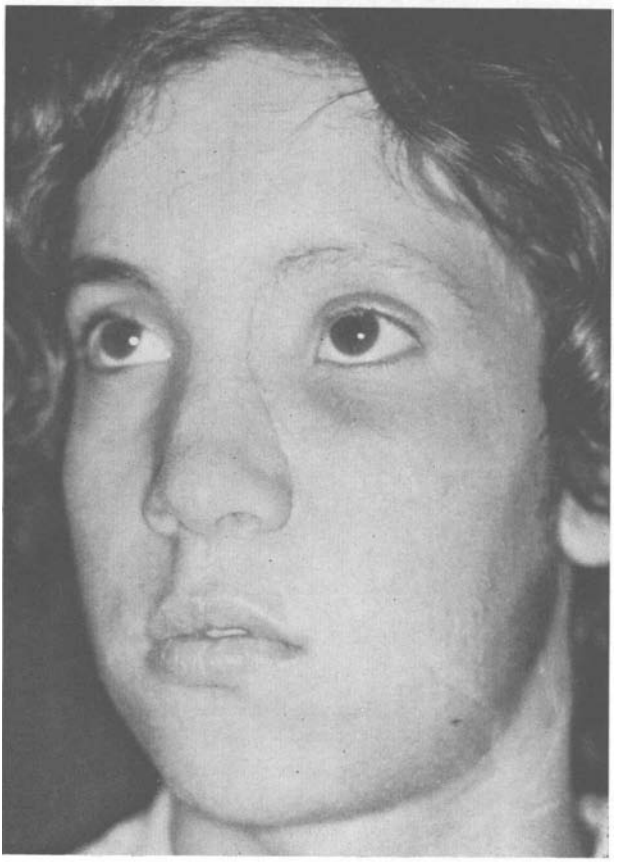


FIG. 10.
Postoperative cosmetic result.

Comment

Olfactory Neuroblastoma is seen mostly in the third and fourth decades. It has never been reported under the age of 9. Both sexes seem to be equally afflicted. McCormack and Harris (1955) further elaborated on the histological classification of Berger as follows:

(1) Tumour with true rosettes—neuroepithelioma.
(2) Tumour with pseudorosettes—neuroblastoma.
(3) Tumour with neurofibril formation, but without rosettes or pseudo-rosettes—neurocytoma.

All the above types have been examined according to their radiosensitivity, spread and survival rate, but no significant difference was demonstrated (Tingwald, 1966).

However, Schenk and Ogura (1973) feel that the above histological features are found only rarely and are of no great importance. We found in our case both rosettes and neurofibrils.

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The tumour has a strong tendency to local expansion and invades the surrounding structures, the paranasal sinuses, the orbit, the meninges and brain, through the cribriform plate. This last route opens the possibility of an ascending infection, and a brain abscess is not uncommon (Robinson and Solitare, 1966). Once in the cranial fossa, tumour cell seeding by means of cerebro-spinal fluid may occur (Riemenschneider and Prior, 1958).

Distant metastases to regional lymph nodes, lungs, liver and other viscera were described (McCormack and Harris, 1955; Seaman, 1951; Fisher, 1955; Becker and Jacox, 1964; Hutler *et al.*, 1959). Clinically the tumour presents as a polypoid mass in the nose, and nasal obstruction and bleeding are the first signs. Anosmia, headache and proptosis follow with the spread of the tumour.

Although the Olfactory Neuroblastoma is considered radiosensitive, it is not radiocurable (Tingwald, 1966), and in our case it was radioresistant.

Hutler *et al.* (1963) in their review of 18 cases, stated that neither radical surgery nor irradiation, nor a combination of both, has been completely effective in controlling this disease. However, a combination of radical surgery and irradiation seems to be more effective than either method alone. Chemotherapy with mechlorethamine hydrochloride and cytophosphamide has been reported effective (Tingwald, 1966) as an adjuvant, especially in advanced inoperable cases. In every case prognosis is guarded and the possibility of local recurrence, intracranial extension or metastasis should be anticipated.

A life-long follow up is recommended (Schenk and Ogura, 1973), as a 5-year survival period in this tumour is meaningless.

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