Olfactory neuroblastoma

By H. Sabeti and A. R. Pashmi (Teheran, Iran)

Neuroblastomas are frequently occurring tumours of infancy and childhood (Russel and Rubenstein, 1963). They arise from adrenal glands, retroperitoneal tissue, sympathetic ganglia and the nasal cavity. Those arising in the nasal cavity form a distinct group.

Case history

A female child of 14 years was admitted to the Ear, Nose and Throat Department of the Amir Aalam Hospital in Teheran on 5 January 1972. She complained of difficulty in breathing through the nose for three months, with severe pain over the right cheek and also had a bloodstained nasal discharge. Breathing through the right nostril was not possible. There was no history of trauma or previous infection.

On clinical examination, a palatal swelling was found to involve half of the hard palate reaching the alveolar margins, and extending to the soft palate posteriorly. It bulged into the buccal cavity (Fig. 1). The right cheek had a soft swelling. The consistency of the palatal swelling was soft and cystic in places. There was proptosis of the right eyeball which was also displaced laterally

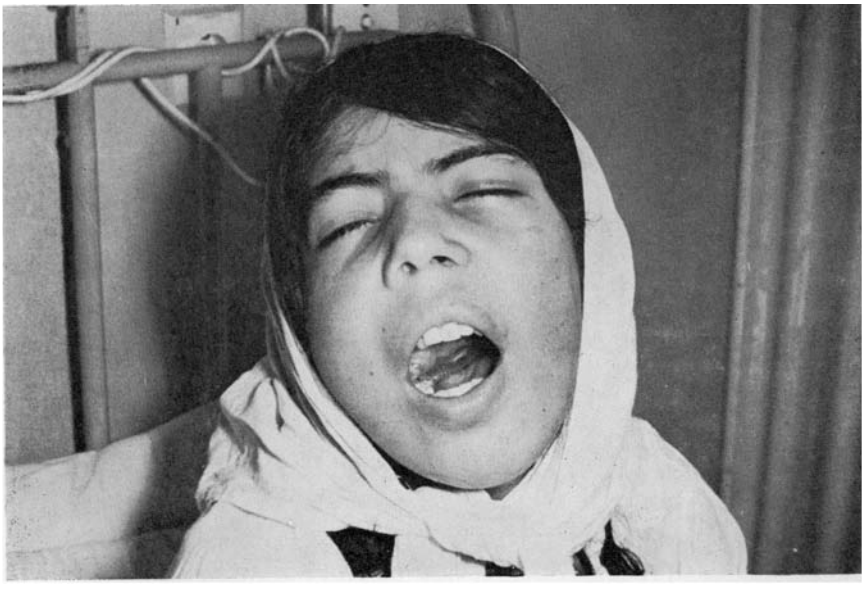


Fig. 1.
Illustrates bulging of the tumour mass into the buccal cavity.

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(Fig. 2). General health was good. The spleen and liver were not palpable, and there was no adenopathy.

Anterior rhinoscopy revealed a collection of pus and a mass blocking the right nostril anteriorly. Posterior rhinoscopy was not possible but palpation of the naso-pharynx revealed a soft friable mass in the roof of the posterior choana.

Results of routine haematological investigations were normal. E.S.R. (Westergren) was 21 mm. in the first hour, blood urea 21 mg./100 ml. and haemoglobin 13 gm. per cent. Examination of the urine did not reveal any abnormality.



FIG. 2.
Illustrates ptosis of the right eye.

Erosion of the medial part of the maxillary sinus was seen on X-ray (Fig. 3).

Roentgengrams of the skull (anterior, posterior and lateral views) showed erosions of the base of the skull (Fig. 4) with haziness of the right maxillary sinus.

A biopsy was taken from the hard palate which resulted in profuse haemorrhage necessitating blood transfusion. Following biopsy, radiotherapy was given for six weeks—a total dose of 6000 Rad. The patient's symptoms improved, the tumour regressed. Surgery could not be done as the patient's guardian sought her discharge against medical advice, on 24 April 1972.

Results of biopsy

Sections from the tumour tissue showed typical neuroblastoma (Figs. 5 and 6): spheroidal cells arranged in sheaths with prominent nuclei and nucleoli with a little cystoplasm and the presence of a tangled mass of neurofibrilis in places. The stroma was vascular. Rosettes could be made out in places.

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Discussion

Olfactory neuroblastoma is a rare tumour. It was reported for the first time in 1924 by Berger and his colleagues (Mendeloff, 1957). Up until 1966, only eighty cases had been reported (Silcox, 1966).

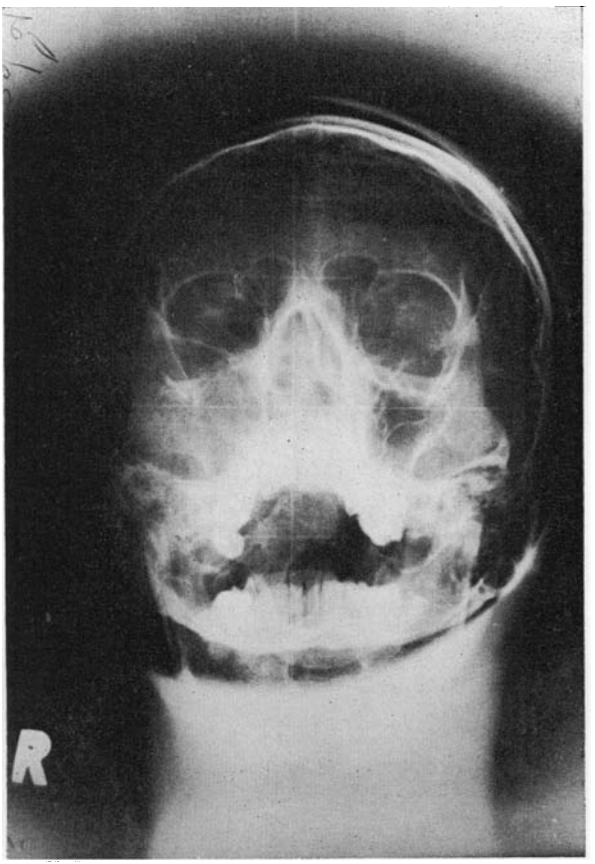


FIG. 3.
Illustrates erosion of the medial aspect of the right maxillary sinus.

The most commonly accepted theory of its histogenesis is that the tumour arises from neuroepithelial cells of the olfactory sense organ. The former component is represented by round or ovoid cells arranged around a central artery (rosettes). Other areas of origin suggested are the olfactory placode, Jacobson's organ, and the sphenopalatine ganglion. The tumour is known to invade neighbouring areas. Our case illustrates this type of invasion very well. Distant metastases have been reported but are very rare. As far as can be judged on clinical and radiological grounds, our case showed absence of metastasis since the tumour responded very well to radiotherapy.

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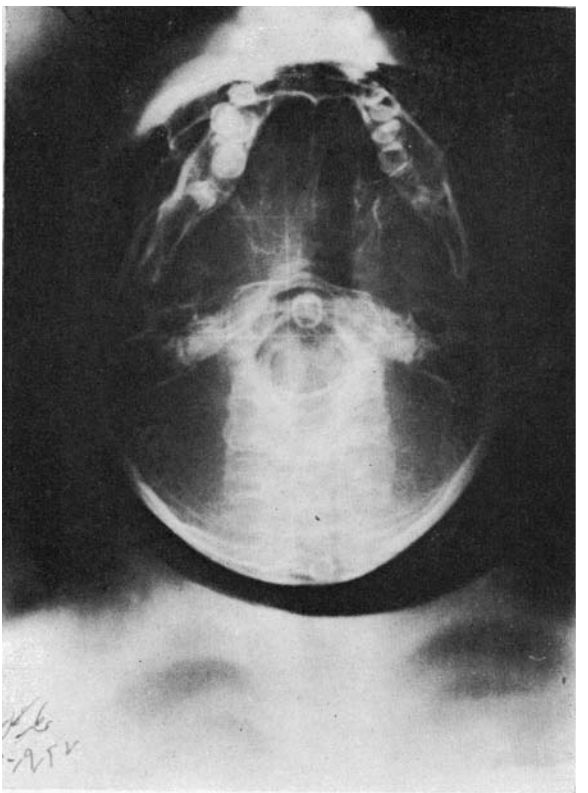


FIG. 4.
X ray of skull showing erosions of the base of the skull.

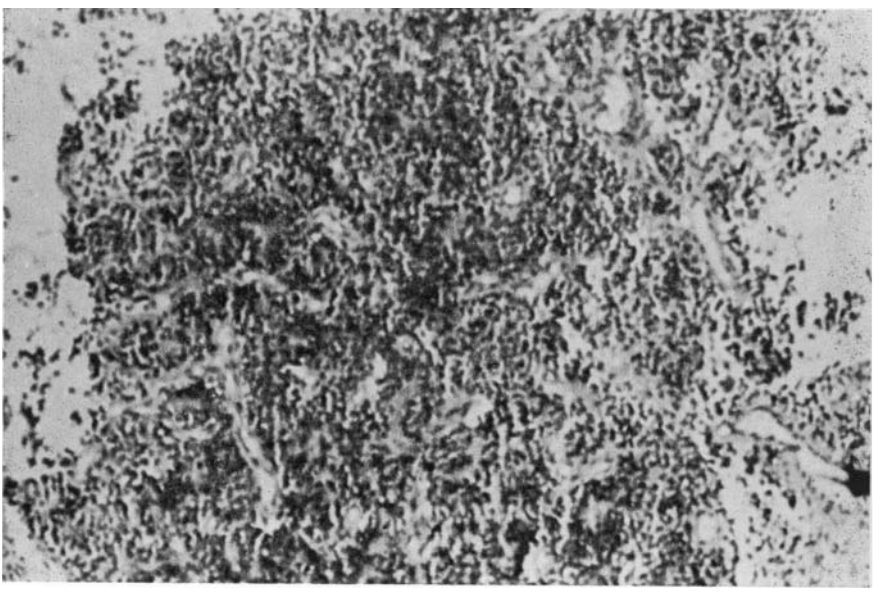


FIG. 5.
Illustrates the typical histological appearance of the tumour stained by H. & E. (× 100).

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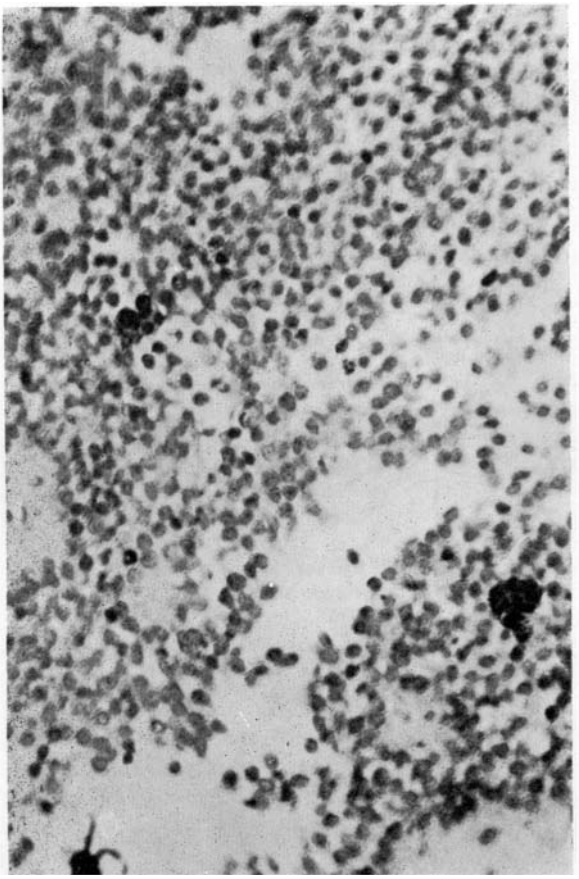


FIG. 6.
Illustrates the same tumour section (×400).

Summary

A case of olfactory neuroblastoma locally infiltrating and involving the palate, orbital cavity, the nose and the maxillary sinuses in a child of 14 years.

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