Olfactory neuroblastoma

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Neuroblastomas constitute one of the most frequent malignant tumours of infancy and childhood (Russel and Rubenstein, 1963). They arise from adrenal gland, retroperitoneal tissue, sympathetic ganglia and nasal cavity. Those arising in the nasal cavity form a distinct group. Olfactory neuroblastoma is a rare tumour. The present communication deals with the report of such a case.

Case report

A female child aged about 13 years (S.K.P., Reg. No. 95697) was admitted under the care of one of us (R.E.V.) on 11 October 1969.

The patient complained of difficulty in breathing through the nose (5 months), swelling of palate and difficulty in swallowing (1 month) and loss of vision in the right eye for last 10 days. Nasal obstruction noticed was first on the right side; later it became bilateral. Loss of vision was gradual. There was no history of trauma.

On clinical examination, the palatal swelling was found to involve almost the whole of it reaching alveolar margins on three sides and extending up to the soft palate posteriorly. It bulged in the buccal cavity (Fig. 1). The right cheek showed a bony swelling obviously due to the expansion of the antero-lateral wall of maxilla. Consistency of the palatal swelling was soft and cystic in places. The right eye ball was pushed outwards and laterally (Fig. 2). Erosion of the medial part of inferior orbital margin was felt on palpation. Anterior rhinoscopy examination revealed collection of pus and a mass blocking nasal cavities on both sides in the posterior part. Posterior rhinoscopy examination was not possible but palpation of nasopharynx revealed a soft friable mass in the roof and at the posterior choana. General health was fairly good. Spleen and liver were not palpable. There was no lymphadenopathy.

Results of routine haematological investigations were normal. ESR (Westergren) was 42 mm./1 hour. The patient belonged to blood groups AB and Rh positive. Examination of urine did not reveal any abnormality. Roentgenograms of skull (anterioposterior and lateral views) showed erosion of the optic foramen (right side) and haziness of maxillary sinuses. External carotid angiography showed that the tumour was fed by the maxillary artery.

After preoperative tracheostomy, lateral rhinotomy approach was made on 24 October 1969. At operation, it was possible to check the extent of the growth: it involved the palate, lateral wall of the nose, orbital cavity and cavity of maxillary sinuses. Complete removal of tumour was not possible. The tumour was partly cystic and highly vascular. Profuse haemorrhage was encountered during its partial removal which was counteracted by transfusion of two units of blood. The patient's symptoms improved. She developed no postoperative complications. Radiotherapy could not be given since the guardian sought discharge against medical advice on 4 December 1969.

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FIG. 2.
Showing ptosis of the right eye ball.



FIG. 1.
Showing bulging of the tumour mass in the buccal cavity.

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Sections from the tumour tissue showed typical neuroblastoma (Figs. 3, 4): spheroidal cells arranged in sheets with prominent nuclei and nucleoli and with little cytoplasm and presence of tangled mass of neurofibrils in places. The stroma was vascular. Rosettes could be made out in places.

Discussion

Olfactory neuroblastoma is a rare tumour. Till 1966, only 80 cases have been recorded (Silcox, 1966). It was reported for the first time in 1924 by Berger and his colleagues (Mendeloff, 1957). The most commonly accepted theory of its histogenesis is that the tumour has its origin from neuroepithelial cells of the olfactory nerve and olfactory epithelial 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 olfactory placode, Jacobson's organ and the sphenopalantine ganglion.

The tumour is known to invade locally neighbouring areas. The present case illustrates this type of invasion very well. Distant metastasis has been reported but is rare. As far as can be judged on clinical and radiological grounds the present case showed absence of metastasis. Since the tumour is known to respond very well to radiotherapy, it was rather unfortunate that the patient could not be given benefit of this treatment.

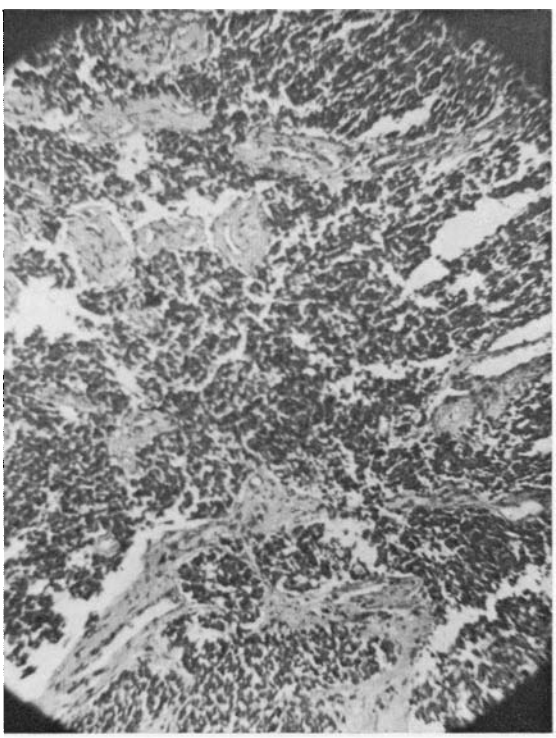


FIG. 3.
Showing the histological appearance of the tumour. H. and E. section × 100.

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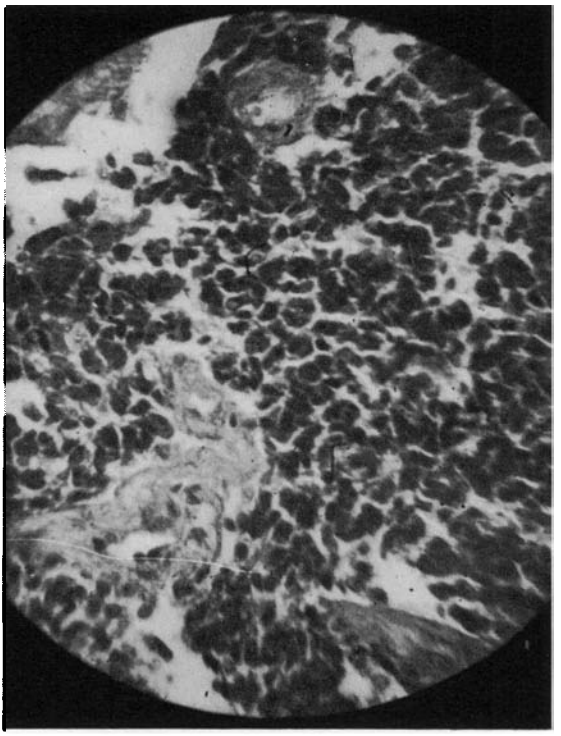


FIG. 4.
Showing the histological appearance of the same tumour at higher magnification. × 400.

Summary

A case of olfactory neuroblastoma locally infiltrating and involving palate, orbital cavity and maxillary sinuses has been reported in a child of 13 years.

Acknowledgement

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