AESTHESIONEUROBLASTOMA OF THE NASAL CAVITY AND MANDIBLE*

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SUMMARY

A case of undifferentiated tumour of the nasopharynx in a young Bantu girl is presented, the origin of which is believed to be an aesthesioneuroblastoma. The literature on this rare neoplasm is reviewed.

Aesthesioneuroblastoma of the nasal cavity is a rare neurogenic tumour of olfactory origin. Reports of this tumour have been infrequent since Berger and Coutard[1] described the first case in 1924, which they called *esthesio-neuroepitheliome olfactif*. By 1961 there were 61 reported cases.[2] The tumour has been variously described as olfactory neuro-epithelial tumour, olfactory neuroblastoma and aesthesioneuroblastoma.

SITE AND ORIGIN

The site of origin remains controversial. They probably arise from neuro-epithelial elements contributed to the olfactory mucosa by the olfactory placode. The stem cell of the olfactory placode is the aesthesioneuroblast, hence the name aesthesioneuroblastoma. Other structures, such as the sphenopalatine ganglion and the remnants of Jacobson's organ have been implicated in their genesis, Herrold[3] succeeded in inducing neuro-epitheliomas in Syrian hamsters by the subcutaneous injection of diethyl-nitrosamine into the interscapular region.

The tumours arise high in the olfactory region of the nose, and may be attached to the cribriform plate or the ethmoidal air cells. The early symptoms are non-specific—unilateral nasal obstruction, epistaxis, anosmia, rhinorrhoea, excessive lacrimation and headache. The tumours are regarded as being of low-grade malignancy but considerable variation in behaviour has been described. They are aggressive growths and may invade the paranasal sinuses, the nasopharynx, the palate, the orbit, the base of the skull and the brain. Metastasis to regional lymph nodes and by the bloodstream has been reported[4-6] and occurred in 14% of the series reported by Hutter *et al.*[7] These authors reported a 5-year survival of 50%, and felt that there was no useful histological guide to prognosis.

MORPHOLOGY

Morphologically these tumours have features in common with neuroblastomas developing in more usual sites, and with certain retinoblastomas. They are not embryonic neoplasms, however, for, unlike retinoblastomas and neuroblastomas of the adrenal gland and sympathetic ganglia, they occur mainly in adolescents and adults (15 - 40 years), but they have been reported over a wide age range: 8 - 79 years.[7]

Macroscopically they are polypoid or fungating, pink to grey fleshy tumours. Histologically they are very cellular neoplasms composed of indifferent neuroblasts.[8] These cells are slightly larger than lymphocytes and consist mainly of an oval or round, mitotically active nucleus with a sharp limiting membrane and loose or finely granular chromatin. The cytoplasm is scanty. The cells are arranged in rosettes or clusters, separated by fibrovascular septa or connective tissue trabeculae. The cells may be set against a fibrillary background, i.e. fibrils extending between the cell clusters, or a web of intercellular fibrils. Obert *et al.*[6] believe that the tumour cannot be diagnosed with certainty unless the fibrils are present. Occasionally the rosettes may resemble those of Flexner, seen typically in retinoblastomas, and they may also resemble the rosettes of adrenal neuroblastomas. They must be differentiated from lymphomas, plasmacytomas and undifferentiated carcinomas.

Neither radiotherapy alone nor surgery alone have proved effective in arresting the growth of this tumour. The best therapeutic approach would seem to be a combination of these. Removal of the ethmoidal extension may necessitate orbital exenteration, according to Obert *et al.*,[6] who also recommend removal of the cribriform plate, with skin-graft coverage.

The following report documents a highly malignant tumour occurring in a young Bantu girl, the origin of which was believed to be an aesthesioneuroblastoma. The author has been unable to find any report of a similar case in a Bantu. The clinical course of this lesion was rapidly fatal—a period of 2 months. The grave prognosis in some of these cases, and the need for urgent treatment, is thus underlined.

CASE REPORT

A Bantu female aged 11 years, presented with left-sided nasal obstruction and toothache of 1 month's duration. A pink fleshy tumour was seen in the left nostril, and the postnasal space was filled with a friable granular tumour. Proptosis of the left eye was present, and a purulent discharge from that eye was noted. The posterior molar of the left side of the mandible was surrounded by tumour which was separated from the main postnasal mass by normal mucosa.

Radiological examination of the sinuses showed destruction of the nasal cavity, septum and left ethmoid, and opacity of the left antrum and orbit. Biopsies from the nose and mandible showed an undifferentiated malignant tumour, the origin of which was believed to be either an aesthesioneuroblastoma or a pharyngeal carcinoma. The sections were seen by Pro-

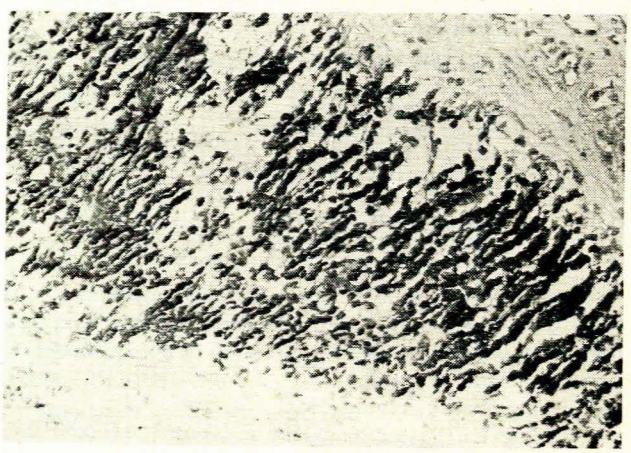

Fig. 1. Medium-power photomicrograph of anaplastic tumour with occasional ganglion cells.

*Date received: 15 March 1971.

fessor L. V. Ackerman, who recommended radiotherapy without delay. However, within a week of admission to hospital the patient's condition deteriorated very rapidly. She was grossly anaemic with a haemoglobin concentration of 6·3 g/100 ml. The red cells showed anisocytosis and some polychromasia, and occasional normoblasts were seen. The tumour enlarged perceptibly in size and distended the soft palate. Respiratory difficulty ensued and a Portex airway tube to facilitate breathing was passed with some difficulty. She died 24 hours later, and was too ill during the 10 days of hospitalization to permit the commencement of radiotherapy.

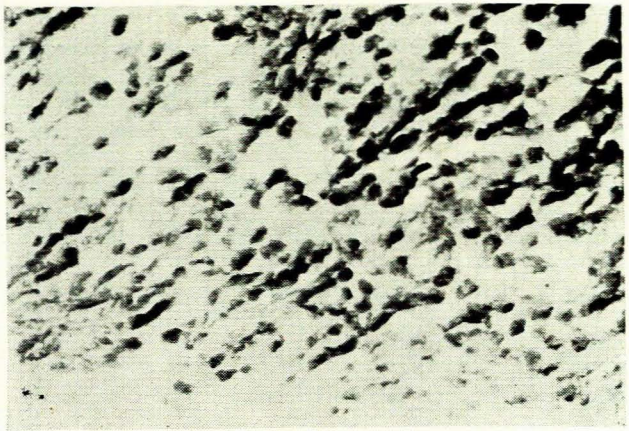

Fig. 2. High-power view of the same tumour.

The author wishes to thank Professor L. V. Ackerman and Dr A. Schmaman of the South African Institute for Medical Research, for interpreting the histology; Dr S. Matic and Dr R. Sack for their assistance in the management of this case; the Superintendent, Baragwanath Hospital, for permission to publish this case and the South African Institute for Medical Research Photographic Unit for the Photomicrographs.

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