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Some rare tumours of the nose and paranasal sinuses

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The tumours of the nose and paranasal sinuses dealt with in this paper are of particular interest for two reasons. First they are rare in this region, and second, they follow an unusual course and are often accompanied by endocranial complications. This paper will comment on the cases of esthesioneuroblastoma, rhabdomyosarcoma and chordoma of the spheno-occipital region. We also comment on a case of sarcoidosis which manifested as a tumour of the dorsi nasi.

ESTHESIONEUROBLASTOMA

The symptomatology of esthesioneuroblastoma in the two patients was almost indentical to classical descriptions in medical literature (frequent epistaxis, difficulty in breathing through the nose, nasal obstruction, hyposmia and headache). The tumours had attacked not only the nose but also one or more paranasal sinuses; thus according to Elkom et al. (1979), they belonged to type B. After radiological and CT scan examination, as well as pathohistological analysis of tissue obtained by biopsy, both patients underwent lateral rhinotomy and the tumours were macroscopically extirpated. The tumorous tissue consisted of agglomerations and focuses of rounded, mainly minute cells. In some places, the cells surrounded small areas of rose-coloured tissue, while in other places they formed dense masses. In some places, there was fine, rose-coloured tissue between the cells.

The younger patient, aged 19, received telecobalt therapy with a tumour dose of 5,300 rads. She has been followed up for 36 months, and there has been no relapse. The second patient did not receive telecobalt therapy. She has been followed up for 14 months.

Most authors recommend a combination of operation and radiotherapy when dealing with type B tumours; Chapman (1981) considers that radiotherapy should be applied as a sandwich therapy, with 2,000 rads preoperatively. Others such as Bailly et al. (1975), Doyle and Paxton (1971) and Scolnik et al. (1966) con-

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sider that radiotherapy should be applied in cases of relapse. We believe that radiotherapy should be applied post-operatively because of the frequency of relapse.

RHABDOMYOSARCOMA

There are three histological entities: a) pleomorphic rhabdomyosarcoma in adults; b) embryological rhabdomyosarcoma, and c) alveolar rhabdomyosarcoma. This is the case of a 63 year old man whose tumour was located on the right alar cartilage. The tumour was extirpated and the pathohistological diagnosis was sarcoma. The recommended post-operative radiotherapy was not carried out to the full but was stopped after 3,500 rads. 12 months later, the patient showed a relapse. The tumour had destroyed most of the right nose, including a part of the alar and triangular cartilages. Frontal tomograms showed that the tumour had spread to the right ethmoidal sinuses and the right maxillary sinus. A lateral rhinotomy was carried out and the pathohistological diagnosis was rhabdomyosarcoma. The tumourous tissue consists of a mass of spindle-shaped polygonal and a symmetrical cells of various sizes. The nuclei are very big and contain large nucleoli. A large number of cells are multinuclear, many of them are grainshaped or look like tadpoles. The cytoplasm of these cells was eosinophilic and granular, but diagonal lines cannot be seen. At the periphery of the tumour there were well-differentiated diagonal striped muscle fibres.

Post-operative radiation therapy was applied to the full tumour dose, as well as chemotherapy. A few months after the chemotherapy, the patient suffered acute leukocytosis, and a needle biopsy of the bone-marrow of the sternum showed that it resulted from acute lymphatic leukemia.

CHORDOMA

This is the case of an 8 year old boy who was hospitalized due to diplopia, difficulty in breathing and mucal nasal drainage from the right nose. Examination showed a tumour on the nasopharynx which had spread through the mesopharynx to the middle of the right tonsil. It was lobular in appearance and covered with an intact mucosa. Ophthalmological examination revealed right convergent concomitant strabism as well as hypermetrophy and differences in protrusion on the papilla n. optici.

The patient was operated on transorally. The extirpated tumour was 3 by 4 cms in size and the pathohistological and electronic diagnosis was chordoma. The electronic microscope revealed a large number of small vacuoles in the cytoplasm of cells and some large vacuoles which deformed their cells into star-like forms. In places the cells surrounded small intercellular spaces containing microvils. Desmosomes could be clearly seen on the surfaces where the cells touched (Figure 1). In addition to the vacuoles, fibrils and free ribosomes in the cytoplasm, there

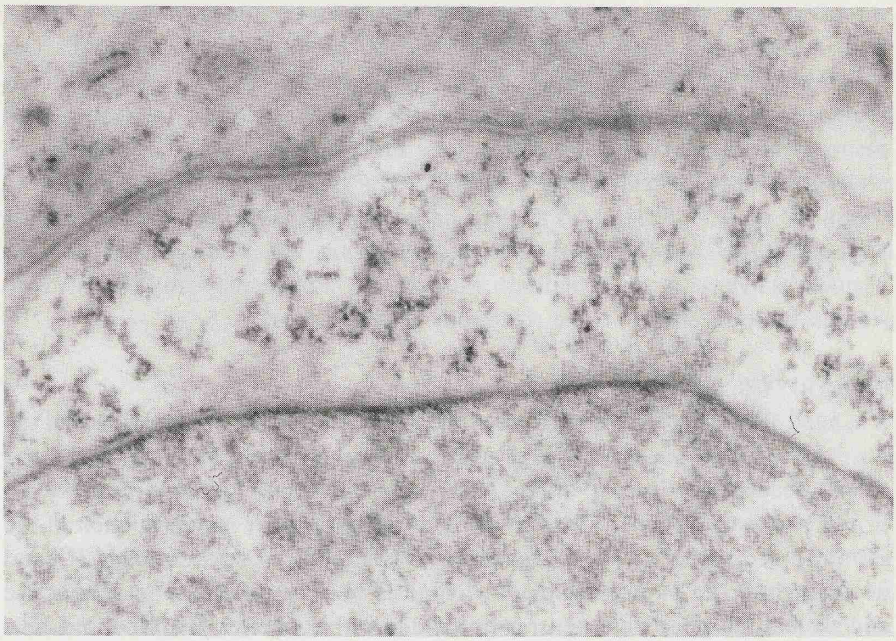

Figure 1. Desmosomes can be clearly seen on the surfac where the cells touch (× 49.700).

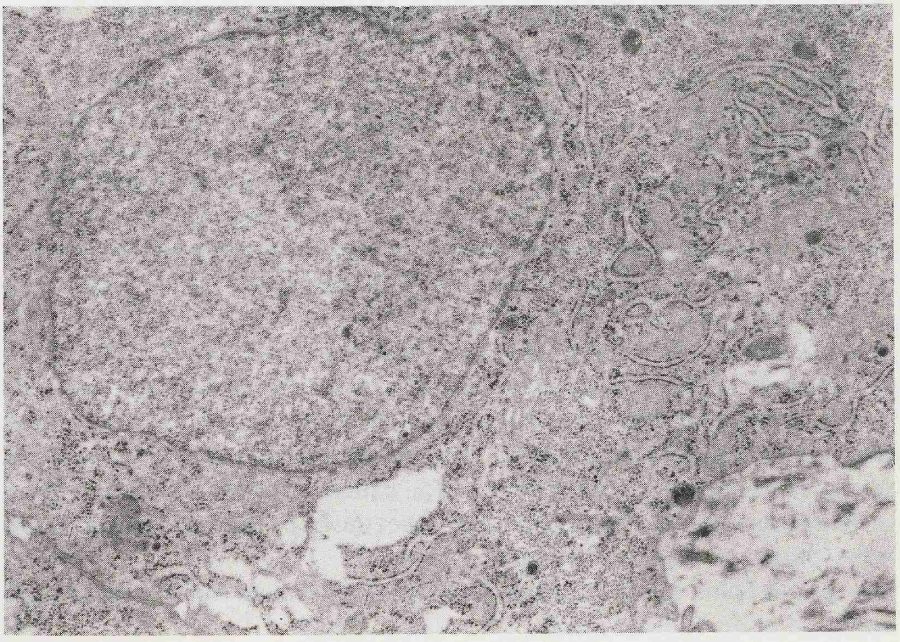

Figure 2. Abundant ergastoplasm in the cytoplasm (× 17.500).

were fairly granulated endoplasmatic reticules and secretory granules (Figure 2). Four months later there was a local relapse of the tumour in the nasopharynx which was extirpated. The patient then went 22 months without discomfort, but then came a range of neurological symptoms including very pronounced central paralysis of the right faciales. There were no signs of the relapse in the nasopharynx. The patient died four months later of endocranial complications.
Campbell et al. (1980) state that eight patients with spheno-occipital chordoma were treated in the Mayo Clinic between 1910 and 1971. The average follow-up period was five years and one month. Surgery and radiotherapy are generally considered the best choice in the treatment of this tumour.

SARCOIDOSIS DORSI NASI

S.I., aged 19, was admitted to hospital because of a tumour at the dorsum of the nose. The tumour was 2 by 2 cms in size; it was smooth and it wasn't painful to the touch. According to the patient, the swelling had appeared four months previously and had gradually increased in size. He had undergone reconstruction of the septum and external pyramid 10 years ago. The tumour was extirpated by ICI and décollement. The pathohistological diagnosis was sarcoidosis. The connective and skeletal muscles were densely imbueded with tubercles of epithelioid cells and rather numerous very large cells, partly of the Langhans type and partly of the foreign-body type. There was no caseosus necrosis, so this was probably a case of sarcoidosis, but an atypical form of tuberculosis cannot be excluded.

COMMENT

Certain rare tumours of the nose and paranasal sinuses, of interest because of their location, their unusual course of their histological picture, were described. Both cases of esthesioneuroblastoma belonged to type B because of the anatomic location of the tumours. One patient received post-operative radiotherapy, which we consider should be applied because of the marked tendency of such tumours to reappear. The second patient merely underwent surgery. Neither patient has yet suffered a relapse.
In the case of a boy suffering from chordoma, it was decided, after the second operation, that radiotherapy would not slow down the process, for there was osseus destruction in the sella turcica and clivus. The patient lived for 26 months after the second operation, which is 50% less that the average survival period in the cases described by Campbell et al. The patient came for treatment with an advanced tumourous process, and we consider this to be the main reason for this relatively short post-operative survival period.
We did not found any description of sarcoidosis dorsi nasi in the most medical literature of the few last years.

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