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NASOPHARYNGEAL CHORDOMA

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Chordoma is a tumour arising from the remnants of the notochord. It is a relatively rare condition and over a period from 1857, when Luschka described the first case, to 1944 only a total of 252 cases were described (Faust, Gilbert and Mudgett 1944). It occurs twice as frequently in males as in females. Cases have been reported in all age groups but most cases present in later life, the mean age for cranial cases being 35 and that for sacro-coccygeal cases being 51 (Willis 1960).

The notochord in the embryo extends from the basiphenoid to the coccyx and it is in these two regions that the chordomas arise most frequently, chordomas of the intervening vertebral column being rare. In the series reported by Faust, Gilbert and Mudgett (1944), 48% were sacro-coccygeal, 37% were cranial and 13% were vertebral. It is at the extremities of the notochord that aberrant residual tissue persists most frequently in adults, and it is therefore not surprising that chordomas have a higher incidence in these two sites.

The chordoma is a locally invasive growth of low grade malignancy. It tends to cause bone destruction and compression of tissues, and erodes rather than infiltrates them. Metastases are rare but not unknown. Of 5 cases described by Willis (1960), 2 of them had blood borne metastases in the viscera. Cases with secondaries in the lymph nodes were also described. These cases were sacrococcygeal growths and most of the cranial growths do not exhibit a tendency to metastasize.

The tumour appears roughly lobulated rounded and circumscribed. It has a gelatinous consistency and there may be cysts, haemorrhages and sometimes calcification. Microscopically there may be great variation in the size of the cells. Some are small and polyhedral, others are large with vacuolated cytoplasm (described by Virchow as physaliphorous cells). These cells may be aggregated together in clusters separated by stromal connective tissue or mucoid matrix.

The clinical features of a chordoma will depend on the site and line of spread of the tumour. They have been well described in Windeyer's review (1959) and later in Ormerod's cases (1960). In the cranial case, which arises in the region of the basi-sphenoid, the growth spreads intracranially at an early stage and death eventually results from compression of vital structures. Apart from invading the cranial cavity the growth sometimes projects into the nasopharynx as well when symptoms of obstruction will be produced. The prominent complaints will therefore comprise of headache and cranial nerve palsies due to intracranial spread; nasal obstruction with discharge, and dysphagia in those with pharyngeal spread. Epistaxis as in nasopharyngeal carcinomas is not common.

Diagnosis is confirmed by radiological findings and by biopsy. Wood and Himadi (1950) in a review of 7 cases of cranial tumours state that the most conspicuous finding in the plain X-ray of the skull is osteolysis of various portions of the sphenoid bone. In all their cases there was destruction of the clivus, dorsum sellae, posterior clinoid process, sellar floor and invasion of the sphenoid sinus. Extension into the nasopharynx and ethmoid sinuses were also found in some of these. Intracranial calcium shadows adjacent to the area of bone destruction were demonstrable in 6 of the 7 cases. It was stated that these shadows could represent either sequestrated bone fragments or actual calcification within the tumour.

Treatment of chordomas is unsatisfactory as the tumour though slow growing is usually well established before diagnosis is made. The site and invasion of bone make complete removal extremely difficult and recurrence after surgery is almost invariable. The tumour is also relatively radioresistant, although some benefit can be derived from a degree of growth restraint (McWhirter and Dott 1955). However, encouraging results have been reported more recently and Windeyer (1959) in his series of 29 cases has shown that radiotherapy has a definite value. In particular, one of his cases who presented in 1941 with a retropharyngeal mass was alive and well in 1959, having received 3 courses of conventional deep X-rays between 1941 - 1942. Ormerod (1960) described a case who received 6000r tumour dose with regression of growth. This patient was treated with another 4000r tumour dose 15 months later

when the tumour recurred; but remained free of clinical recurrence in the nasopharynx 4 years after this second course of treatment.

The following is a case report of a cranial chordoma presenting in the nasopharynx. The patient was seen at the E.N.T. Department and treated at the Radiotherapy Department, General Hospital, Singapore.

G.G.C. a male Chinese aged 42 presented in the E.N.T. Department, General Hospital on 5.2.58. His main complaints were headache off and on for about 4 months associated with numbness of the right side of the face. There was also blurring of vision in the right eye and right nasal obstruction. On examination, at that time, there were no demonstrable cranial nerve lesions and the cervical lymph nodes were not palpable. Post-nasal space examination revealed a smooth rounded tumour occupying the right choana.

X-ray of sinuses and base of skull on 2.5.58 showed both maxillary sinuses hazy, the right more than the left. There was a mass on the right side in the base of skull view with possible erosion in the region of the apex petrous temporal bone.

Two biopsies of the growth were taken but were reported as being not conclusive but suggestive of malignancy.

In view of these findings, the case was regarded as a nasopharyngeal carcinoma (which is the commonest malignant tumour of the post-nasal space among the Chinese in this part of the world). On 24.6.58 he was started on deep X-ray therapy (250 K.V., H.V.L. of 3.5 mms. Cu.) and a total tumour dose of 5292r was given over a period of 5½ weeks. When examined on 13.8.58, two weeks after completion of treatment, the headache was better but he still complained of nasal obstruction and some blurring of vision, and there was no diminution in the size of the growth.

About a month after the irradiation, on 27.9.58, there was still no appreciable regression of the tumour and it was felt that there must be some explanation for the peculiar radioresistance of the growth. (Nasopharyngeal carcinomas, in our experience, had almost invariably been extremely radiosensitive). A further biopsy of the post-nasal space was carried out on 9.6.60, but this again did not reveal any positive results. The patient continued to be followed-up at regular intervals and was treated symptomatically.

However, on 17.10.61 when he came up a biopsy of the tumour was done again and the report was as follows:—

"Tissue from roof of right choana —
Section shows a neoplasm composed of irregular groups of pale irregular and occasionally vacuolated cells in a markedly mucinous matrix. The structure is consistent with a chordoma".

The slides were reviewed by the Pathologist and the diagnosis of chordoma was confirmed.

The patient's symptoms improved and in fact after the last biopsy he did not present with any complaints apart from blurring of vision in the right eye.

An X-ray of the skull on 11.1.63, however, showed a large soft tissue mass occuying the right nasal cavity. The medial wall of the right maxillary sinus as well as the ethmoidal sinuses appeared to be eroded. The growth had also involved the sphenoid sinus and caused destruction of the sella turcica. The clivus was also partially eroded and there was calcification just above the pituitary fossa (see Figs. 1 & 2).

When seen on 23.4.63 examination of the post-nasal space showed that there was still a bluish fleshy growth in the right choana. This was much smaller than before. In view of this, surgery was suggested to the patient but this he refused as he was relatively symptom-free. On 22.5.63, fundi examination showed optic atrophy on the right side. Other cranial nerves were normal. He is still on regular follow-up and was last seen on 12.6.63.

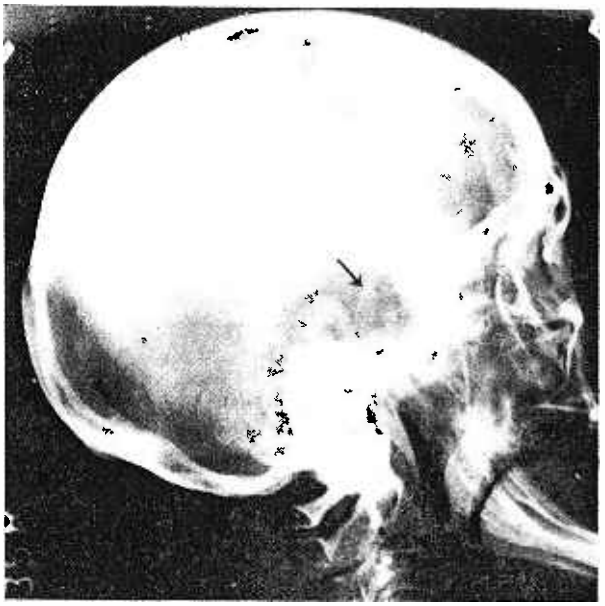

Fig. 1. Arrow points to area of calcification above the destroyed sella turcica.

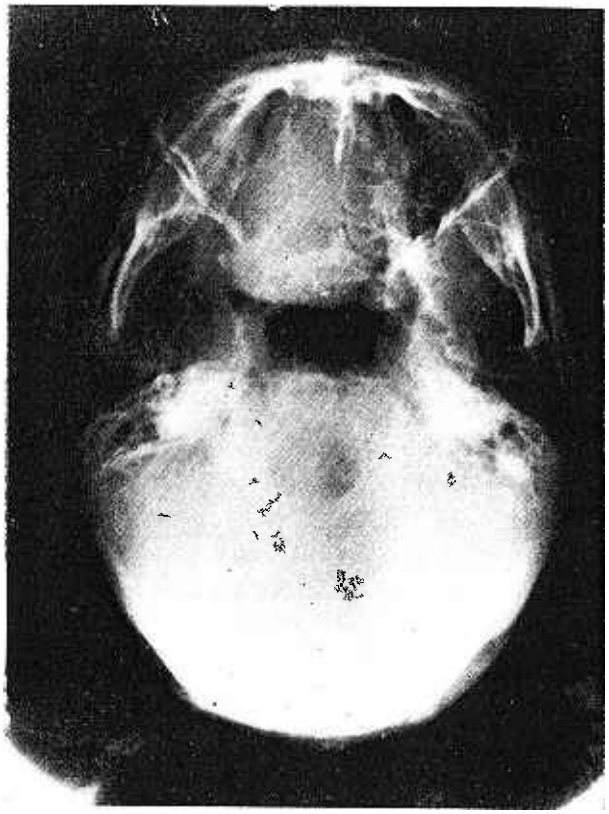

Fig. 2. This shows the soft tissue mass in the region of the right nasal fossa which has eroded the medial wall of the maxillary antrum.

DISCUSSION

As stated above, in Singapore, most of the malignant tumours of the nasopharynx in Chinese have been nasopharyngeal carcinomas. The diagnosis in the above case had to be reviewed because of the slow course of the disease and the atypical response to radiotherapy. The earlier X-rays of the skull were not conclusive, but the later films may be regarded as very suggestive of a chordoma.

The prognosis in chordoma is almost uniformly bad. Complete eradication of the growth either by surgery or radiotherapy is practically impossible. However, owing to the slow rate of growth long periods of survival may be achieved. This is said to be the case in many cases of sacro-coccygeal chordoma. In cranial cases, owing to the spread intracranially, survivals of more than 5 years are rare (McWhirter and Dott 1955). Of the above case, it may be said that satisfactory growth restraint has been achieved with radiotherapy. There is still a residual tumour for which palliative surgery or further irradiation can be given, should symptoms arise. From the report of Windeyer's cases (1959) it would appear that higher doses of radiation might be necessary for chordomas to achieve satisfactory results, and also that supervoltage therapy would have definite advantages. However, the danger of over-irradiating the central nervous system, with consequent damage to the brain stem or cervical cord must be borne in mind in the radiotherapy of this region.

SUMMARY

(1) The pathology and clinical features of chordoma are reviewed and reference is made to the radiological findings in cranial chordomas.

(2) A brief discussion on the treatment follows.

(3) A case of chordoma presenting in the nasopharynx is described. This is discussed with reference to prognosis and future management.

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