# PARAPHARYNGEAL CHORDOMA

## A case report

P.J. DAMATO

M.D., D.L.O., F.R.C.S.

J. PACE BALZAN

M.D., F.R.C.S.

The parapharyngeal gutter is a triangular space bounded medially by the buccopharyngeal fascia and posteriorly by the pre-vertebral musculature, while anterolaterally it is roofed over by the sternomastoid, strap muscles and the encircling layer of the deep cervical fascia. It extends from the base of the skull into the superior mediastinum. It contains the carotid sheath, the last four cranial nerves and the deep cervical and retropharyngeal lymph nodes.

The commonest swelling in this space follows enlargement of the deep cervical nodes. Neurofibroma, chemodectoma and aneurysm of the carotid artery are occasionally found in this region.

The purpose of this paper is to report an unusual case of a chordoma arising in the region of the foramen magnum and extending laterally to present as a parapharyngeal tumour.

### Case Report

The patient, a middle aged male, presented with a recent aggravation of a 10 month history of periodic neuralgic pain in the left side of the face and head radiating to temple and throat. He also complained of left sided deafness, and difficulty in swallowing both liquids and solids. His voice was normal.

Positive physical findings included:

(i) Fullness of the left side of the neck below the angle of the mandible, continuous medially with a parapharyngeal mass which had displaced the tonsil across the midline. It extended upwards into the nasopharynx and distally to the level of the hyoid. It was smooth, firm and non-pulsatile. There were no palpable nodes. (ii) Paralysis of the last four cranial nerves. (iii) Left secretory otitis media.

### Investigations

1. Audiogram: Lt. conductive deafness averaging 30 — 40 db over the speech frequencies. 2. Haematology: normal blood picture. 3. Radiology: (a) Lateral soft tissue views of the neck showed a degree of widening of the retropharyngeal space. (b) Submentovertical views showed widening of the left jugular foramen. (Fig. I). (c) Tomography confirmed the above findings and showed some irregularity in the anterior margin of the foramen magnum suggestive of early bone destruction. (d) On angiography the carotid arteries were splayed out by the tumour, but no abnormal tumour circulation was demonstrated. (e) Chest Xray was normal.

The mass was removed through a lateral cervical approach. It was a large multilocular cystic tumour filled with jelly-like material. It appeared to be well encapsulated and extended caudally to the level of the hyoid. The internal carotid artery was postero-lateral to the tumour; the external carotid disappeared deep to it. The hypoglossal nerve was stretched across it.

The lower half of the tumour was readily dissected away from the surrounding tissues with preservation of the hypoglossal nerve. More cranially the mass was followed to the base of the skull, above and

deep to the transverse process of the atlas where the tumour broke open. The remaining cavity, about 1 cm in diameter, was drained, its mucoid contents being sucked out. There did not appear to be any connection between the tumour and the pharynx or subarachnoid space.

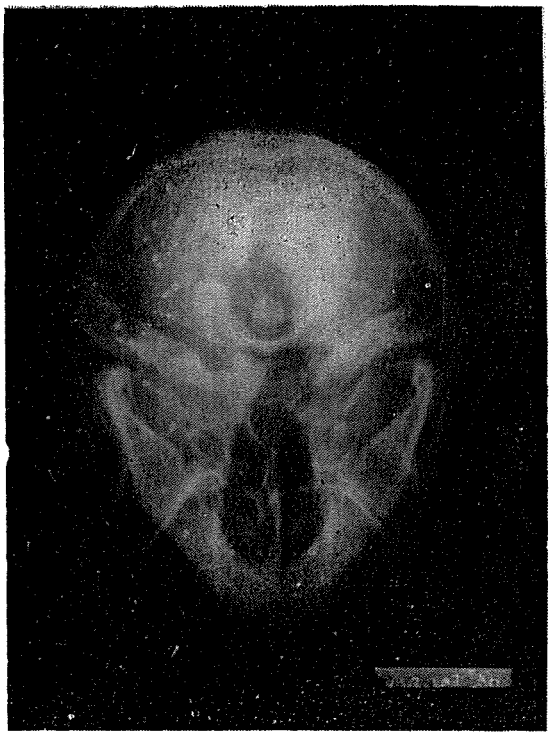

**Fig. 1.**

Postoperative recovery was uneventful and marked by immediate relief from pain.

*Histology report:* Section shows large vacuolated cells and in some areas syncytial strands of cells all within a matrix of mucus. In the more cellular areas pleomorphism is present. This picture is suggestive of Chordoma.

## Discussion

Chordomas are rare tumours constituting less than 1% of all neoplasms affecting the Central Nervous System (Poppins and King 1952). They are some of the few malignant tumours originating from vestigial embryonic tissue which retain primitive histological features. They are believed to arise from remnants of the notochord, the primitive axial structure first found in Amphioxus and Tunicata and around which the vertebral column is formed in higher vertebrates (Rewell 1963). What precipitates malignant change in these remnants is not known, but trauma is believed to play a part. (Utne and Pugh 1955).

The theory that these tumours originate from notochordal rests is strengthened by the following facts: (i) In the cranial region these tumours are never found anterior to the pituitary fossa. In man, the cranial end of the developing notochord enters the basi-occiput for a short distance until it comes into contact with the endothelium of the primitive pharynx caudal to Rathke's pouch, within the body of the sphenoid, caudal to the pituitary fossa. (ii) Notochordal rests have been demonstrated at the three main sites of origin of cranio – cervical chordomas viz. the posterior wall of the pituitary fossa, the posterior pharyngeal wall, and the anterior margin of the foramen magnum. (Wright 1967).

These tumours may arise at any point in the cranio – vertebral axis, but are most common in the sacrococcygeal region and base of skull. The higher incidence of chordomas at both extremes of the axial skeleton may be explained by the greater developmental activity in these regions consistent with the development of head and tail folds.

The occasional finding of chordomas at sites away from the axial skeleton is not so readily explained. Cases have been described in the maxillary and frontal sinuses. (Pastor *et al* 1949; Adams 1945). The possibility exists that in these situations the tumours arise from persistent foci of chordomatous tissue that has been displaced more widely than usual. The subject is however obscure and the possibility of mis-diagnosis cannot be ruled out owing to the difficulty of distinguishing them histologically from the more common tumours in this region, like mucin – forming adenocarcinoma.

The histological appearance of chordoma may vary widely but the characteristic feature is the "Physaliphorous cells" of Virchow — cells blown up by excessive production of mucus. These cells gradually

lose their cell outline and in parts appear as a vacuolated syncytium with a few cells surrounded by mucus. The typical chordoma is a slow – growing tumour which extends into the soft tissues pushing them aside with erosion rather than infiltration. Bone at the site of origin may show extensive absorption. Metastases, which may occur in some 10% of cases, are found in lung, liver, lymph nodes and subcutaneous tissues. These are less common with cranio-cervical chordomas possibly because of earlier presentation and an earlier fatal outcome of tumours at this site. Cranio – cervical chordomas usually present about the 4th decade i.e. earlier than chordomas at other sites. These is no appreciable difference in sex incidence.

Symptomatology varies with the site of origin of the tumour, but pain is the earliest and most common presenting symptom and may be due to erosion of bone or cartilage or involvement of nerve roots. Visual disturbances with loss of visual acuity, diplopia and limitation of visual fields are characteristic of spheno-occipital chordomas, while extensions into the hypothalamic area present with endocrinal dysfunction. Cranial nerve involvement occurs progressively with lateral extension of the originally midline tumour.

Treatment is usually surgical and incomplete because of bone involvement at the time of diagnosis. The commonest approach for cranio – cervical chordomas is the lateral cervical or transpalatal. High dosage radiotherapy has occasionally been used to control tumour residue in bone and recurrences. (Ormerod, 1960; Boyle and Frank 1966). Chordomas are not completely radio-resistent, and the chordomas of childhood are more sensitive than those seen in the adult.

Prognosis is poor, the average survival of cranial chordomas being 3 years from the onset of symptoms. Cases have, however, been recorded of patients surviving in good health for much longer periods following repeated excision of tumour recurrences.

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