Double Midline Intracranial Tumors of Vestigial Origin: Contiguous Intrasaral Chordoma and Suprasellar Craniopharyngioma

Case Report*

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This report describes the simultaneous occurrence of an intrasellar chordoma and a suprasellar craniopharyngioma in the same patient, a combination which to our knowledge has not previously been recorded. The diagnostic problems posed by the coincidence of 2 superimposed midline intracranial tumors, and the oncologic questions raised by the concomitant development of 2 different neoplasms of vestigial origin constitute the main interest of this case.

Case Report

History. A 53-year-old real estate broker (B.S., Hosp. #800-89-67) developed low back pain radiating to the left hip and leg in February, 1968. Examination at an outside hospital revealed a left foot drop with hypalgesia and muscle weakness in the 5th lumbar nerve root distribution, typical of intervertebral disc protrusion at L4-L5. This was confirmed by myelography. There was also an incomplete left 3rd nerve palsy. On March 4, 1968, laminectomy at L4-L5 on the left revealed a protruding disc at that level.

Several days later the patient developed prolonged mental confusion. Electroencephalography revealed diffuse slowing without focal abnormality. A roentgenogram of the skull demonstrated marked erosion of the sella turcica with extensive destruction of its floor, consistent with an intrasellar mass (Fig. 1). Pneumoencephalography confirmed the presence of this large mass extending apparently beyond the sella. The 3rd ventricle was not visualized.

Operation. On March 18, craniotomy disclosed a large encapsulated intrasellar tumor which contained numerous small cysts. Some of these were evacuated, and a large portion of the mass removed, the microscopic structure of which was reported to be that of a chordoma.

Course. Following operation the partial left 3rd nerve lesion increased to complete palsy. The patient also continued to show marked incoordination. Spinal puncture revealed increased intracranial pressure, and on April 24, 1968, a ventriculo-atrial shunt was performed. Radiotherapy (6000 r) was also directed to the sella. The ataxia and 3rd-nerve palsy progressively improved over the next 6 months. The ataxia then returned and, in December, 1968, an electroencephalogram showed spotty focal abnormality and slow-wave dysfunction. A pneumoencephalogram failed to fill the anterior portion of the 3rd ventricle; this was attributed to extension of the intrasellar mass. A second ventriculooatrial shunt was performed on the opposite side, followed by clinical relief provided both shunts were kept patent by pumping.

In May, 1964, however, ataxia and progressive disorientation returned, culminating in a marked confusion. Examination at the Palo Alto-Stanford Hospital elicited a complete left-sided ptosis, diminished visual acuity, normal visual fields, no papilledema, and no other abnormal physical signs. Laboratory investigations of blood, electrolytes and urine were normal. The cerebrospinal fluid obtained at pneumoencephalography was clear and colorless; it contained 10 white blood cells (5 neutrophils and 5 lymphocytes) per c. mm., 12 red blood cells per c. mm. and 172 mg. of protein per 100 ml. A millipore filter from the fluid was positive for tumor cells. A roentgenogram of the skull showed gross enlargement of the sella, with erosion of its floor, and thinning with posterior displacement of the dorsum; the anterior clinoïd processes were, however, present. Pneumoencephalography and ventriculography demonstrated a large mass occupying most of the 3rd ventricle (Fig. 2). A lateral view disclosed that this mass was...
completely surrounded by air, and thus quite separate from the intrasellar mass (Fig. 3).

Second Operation. On June 2, 1964, a second craniotomy was performed, and the 3rd ventricle approached transventricularly. A biopsy of the mass from the 3rd ventricle revealed a tumor which, microscopically, was entirely different from the previous biopsy and whose histological appearances were typical of craniopharyngioma.

Course. The patient did not recover from the operation, remained comatose, and expired on August 10, 1964, 20 months after the onset of his symptoms.

Post-Mortem Examination. (Autopsy #4-319). The major findings outside the central nervous system consisted in an acute bilateral staphylococcal bronchopneumonia accompanied by septicemia.

The brain weighed 1300 grams. The pial surface showed slight yellowish discoloration indicative of old leptomeningeal hemorrhage. On retracting the frontal lobes, a greenish-grey gelatinous suprasellar mass, located and focally calcified, with a smooth inferior surface was found above the optic chiasma, widening the interpeduncular fossa. The intact pituitary stalk was noted to be in its normal position. The hypophysis was raised slightly above the diaphragm sellae and compressed by a second underlying intrasellar neoplasm unconnected with the first. This neoplasm was partly greyish-pink and gelatinous, partly hemorrhagic. It had eroded the enlarged sella, which measured 3 cm. in diameter (Fig. 4). It was well defined and partly encapsulated, but had extended into the sphenoidal and the right maxillary sinuses. The walls of the sella were softened throughout and could be cut with scissors.

Mid-sagittal section of the fixed brain demonstrated replacement of the posterior two-thirds of the 3rd ventricle by the suprasellar tumor, which was well circumscribed, partly cystic and partly solid, with a greyish gelatinous cut surface punctuated by yellowish foci (Fig. 5). The cystic areas were intersected by greyish-white strands of fibrosis. Anteriorly, the tumor was firmly tethered to the optic chiasm and optic nerves. Posteriorly, it was separate from the pineal, which was easily identified. Subsequent coronal sections demonstrated bilateral displacement of the thalami by the centrally located neoplasm, which also compressed the left substantia nigra and cerebral peduncle.

The left calcarine cortex was the site of an extensive old hemorrhagic infarct, and symmetrical foci of old infarction were also noted to involve both halves of the tegmentum in the more rostral portion of the pons. These lesions were interpreted as secondary to a previous episode of tentorial herniation.

Microscopic Study. The previous surgical biopsies were reviewed, and sections from various blocks of the 2 tumors found at autopsy, as well as from the adjacent

Fig. 2. Pneumoencephalogram. Large midline mass replacing most of the third ventricle and invaginating its floor.

Fig. 3. Pneumoencephalogram. Lateral view. Air in the subarachnoid space clearly separates the suprasellar from the intrasellar mass. Note also air filling only the posterior portion of the third ventricle (arrow).

Fig. 4. Autopsy specimen. Superior view of sella turcica, almost completely occupied by intrasellar chor- doma. AC: Anterior clinoid process; DS: Dorsum sellae; ICA: Internal carotid artery; PS: Pituitary stalk.