CASE REPORTS AND TECHNICAL NOTES

INFUNDIBULOMA

A CASE REPORT WITH A BRIEF REVIEW OF THE LITERATURE

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The purpose of this paper is to report the fifth case of infundibuloma recorded in
the literature since 1942 when Globus first studied and described a hitherto un-
known tumour of the infundibular region. Prior to this date, these neoplasms may
have been placed in groups to which they bore some resemblance, but the work of
Globus showed they belonged to a category which was quite distinct and previously
unrecognised. In the case presented below the patient was the youngest of any that
has so far been reported and we feel that the tumour was present since birth.

REVIEW OF THE LITERATURE

Of the many tumours in the region of the 3rd ventricle, which have been exhaus-
tively studied, the infundibuloma first described by Globus presents certain char-
acteristic features which make it a distinct and rare entity. He recognised that these
tumours contained structures which occur only in the normal infundibular region,
thereby providing landmarks for purposes of identification. In the normal tuber
cinereum there is a system of blood vessels which had been previously termed the
hypophysio-portal system by Popa and Fielding. These vessels take origin from
the sinusoids of the buccal portion of the hypophysis and from the capillaries of
the neural portion, ascending through the stalk to the region of the infundibular re-
cess of the floor of the 3rd ventricle, where they break up into a secondary capillary
reticulum. In the course of their ascent, they acquire "glial" sleeves as far as the floor
of the infundibular recess; at this point they lose these glial coverings and then con-
tinue as a capillary network. It is the presence of these unusual vessels that consti-
tutes a "landmark" in the infundibular region. Globus also investigated the embry-
ological and comparative anatomical structures analogous to this hypophysio-portal
system since, according to Tilney, the saccus vasculosus, which is represented by
the dorsal saccular surface of an infundibular process, might in fact be the phylo-
genetic precursor of these bizarre tumours. Globus, however, was unable to come
to any definite conclusion in this regard.

He reported 2 cases. Both patients presented typical signs of increased intra-
cranial pressure, and both died within 48 hours after craniotomy. Necropsy was per-
formed. Histological examination of the tumours revealed mainly elongated unipolar
or bipolar cells, similar to the pituicytes found in the posterior lobe of the pituitary.
Often these cells were aggregated into tracts of pseudo-fibres, and for this reason
some investigators may have considered these tumours neurinomata or polar
spongioblastomata. There were vascular channels described as "vascular lakes," and
numerous vessels that were similar to those of the hypophysio-portal system. Among
the cells were numerous bodies bearing considerable resemblance to the colloid-like bodies found in the normal neurohypophysial region around the hypophyseal portal vessels.

Papez and Ecker reported a case of infundibuloma associated with precocious puberty in an 8-year-old boy. Histological examination, performed after the boy had died on the 3rd postoperative day, revealed structures similar to those described by Globus, and the pathological diagnosis was in fact confirmed by the latter investigator.

Fine and Goldfarb described a case of a 6-year-old boy, dying within 48 hours of craniotomy. Necropsy revealed a neoplasm similar in type to that described by Globus, with the characteristic “glial-sleeved” blood vessels and typical cellular elements.

It is interesting to note that in all of the above cases the patients showed unusual eye signs, including pathological movements and disturbances of vision, as did our patient.

CASE REPORT

Pat Mc., a boy 1 year of age, had been in reasonably good health until 3 months prior to admission, when he commenced vomiting and began to lose weight steadily. He vomited every feeding, sometimes in a projectile fashion, but retained a small portion. He was an extremely placid infant, but became rather irritable when moved suddenly. During this time he had been running a low grade of fever, associated with a slight cough. He had never been cyanosed or had convulsions, but had been constipated since birth. He weighed 8 lbs. 8 oz. at birth and on admission weighed 18 lbs. Three months before admission he had begun to support himself on his legs, but gradually ceased to do this, and in fact resented being forced to sit upright. He was less active than he used to be although he had never been a very active baby. He seemed to be in pain at times, rubbing the back of his neck and holding his head to the right. He had had rotatory eye movements since shortly after birth, and sometimes he made curious movements of his hands, the upper part of the arm being adducted, the elbow flexed, and the hands slowly pronating and supinating.

Birth History. He was a full term normal delivery, and had not been breast fed. He sat up at 6 months and cut his first tooth at 2 months.

Examination. He was a small baby lying very still, with an obviously enlarged head. The circumference of the skull measured 19½ inches. The anterior fontanelle was widely patent and tense.

He had marked rotatory nystagmus, worse on the right than on the left. There was a slight degree of rigidity of the neck and limitation of straight leg-raising. Muscle tone was good. Percussion of the skull revealed a “cracked-pot” sound over the temporal areas. A fluid thrill was also present in this region. No vascular bruits were heard. Reflexes were brisk; the right knee jerk was slightly brisker than the left. Bilateral Babinski response was present.

The right optic disc was pink and the margins were blurred with early papilloedema of 1 to 2 D. The left disc showed late and severe optic atrophy. Both pupils reacted very poorly to light.

Course. He continued vomiting, and parenteral fluids were administered. Ventriculography revealed a marked hydrocephalus. Both lateral ventricles were greatly dilated. There was no filling of the 3rd ventricle, but air did pass into the aqueduct and into the 4th ventricle, which was normal in size, shape and position. Destruction of the sella was noted. A rounded mass was seen elevating the tip of the left frontal horn and compressing the tip of the left temporal horn, suggesting an infundibular tumour with upward extension into the left frontal and temporal lobes.

Operation. Decompression of the ventricular system by needle drainage was done, affording adequate visualization of the anterior and middle fossae. On elevating the left temporal lobe