COLLOID CYST OF THE THIRD VENTRICLE ASSOCIATED WITH CONGENITAL CYSTIC KIDNEYS *

BY E. WYN JONES AND DOUGLAS H. COLLINS, LIVERPOOL

While true neoplasms in the region of the third ventricle are of fairly common occurrence, colloid cysts within its cavity are somewhat infrequently met with. The first two cases in this country were published by Batten and Collier (1899) and Mott and Barratt (1899); these were followed by Kinnier Wilson's case in 1906. The first series of cases was collected by Hall (1913), who found seven cases in the literature and added two of his own. Twenty years later, Zimmerman and German (1938) reviewed all the recorded cases, 28 in number, and added two more, one of which was successfully operated on. Dandy (1934), in his recent book on benign tumours of this area, records operations on five cases of colloid cyst. The cyst wall was removed completely in all and there was only one fatality—a month after the operation—due to blocking of the aqueduct of Sylvius. Cysts in this region are not always of choroid plexus or ependymal origin. In a number of reported cases they have been epidermoid or parasitic in nature; others are stated to be derived from the hypophysis. One case was considered by the author to be a degenerated sarcoma—myxosarcoma, while other cases appear to have been cystic dilatations of the cavum septi pellucidi—a condition that has been described by Dandy (1931). Byrom and Russell (1932) considered that of all the reported cysts in this region only 20 were of the ependymal or choroid plexus type, and that some of these were inadequately described histologically.

The following case is recorded because of the rarity of colloid cysts of the third ventricle, the difficulty of diagnosis and the association of the condition with congenital cystic disease of the kidneys. It is not proposed to review the literature on the subject, as this has been done recently by Zimmerman and German. Tumours in the region of the third ventricle have been extensively dealt with by Fulton and Bailey (1929), who include a case of colloid cyst in their series. Both of these articles are accompanied by a comprehensive bibliography.

* From Liverpool Royal Infirmary and Pathological Department, University of Liverpool.
REPORT OF PERSONAL CASE

Clinical Findings.—The patient, a schoolboy, age 16, was referred to one of us by Dr. R. G. McWhinney after two days' illness. His previous history contained nothing relevant apart from the history of a slight knock on the head two days prior to admission. He had always been a healthy boy and the family history was clear.

It was stated by his parents that he was perfectly well until about 48 hours before coming to the hospital. He then began to complain of severe headache, which failed to respond to the usual remedies. Within a few hours nausea was complained of, and vomiting soon became a troublesome feature. By the following day he was stuporose, and his attention was only attracted with difficulty.

When admitted to hospital the next day, the patient was comatose, but he resented actively the process of examination. His attitude was that of cerebral irritation, and a moderate degree of photophobia was present. Although of rather slight build, he was of average muscular development, and early secondary sexual characteristics had appeared. The pulse rate was 60 per minute, and the temperature was 97° F.; breathing was quiet and the respiratory rate was 24. There was no rash, cyanosis, anaemia, clubbing of the fingers or jaundice. Proptosis and exophthalmos were not observed.

Adequate neurological examination was impossible owing to complete lack of cooperation on the part of the patient. Slight bilateral papilledema was found. There were no retinal hemorrhages or exudates. The pupils were somewhat dilated and their reaction to light was sluggish: there was some degree of paralysis of associated ocular movement, but the attempt to analyse it was not very satisfactory. The main ocular paresis appeared to affect both internal recti. There was no weakness of the external recti. The corneal reflexes were present and equal. The usual test for cerebellar dysfunction could not be carried out. The limbs were neither spastic nor hypotonic, while the knee and ankle jerks were sluggish and the plantar responses were bilaterally extensor. Kernig's and Brudzinski's signs were negative, and there was no head retraction. There seemed to be a generalized hyperalgesia. Clinical examination revealed no gross abnormality of the heart and lungs. The lower pole of the right kidney was easily palpable; the liver, spleen and left kidney were not felt. The patient had urinary incontinence, and on catheterization only a few cubic centimetres of urine were obtained. This was found to be loaded with albumin, but no sugar or acetone was present.

A tentative diagnosis of subarachnoid hemorrhage was made. Lumbar puncture revealed spinal fluid under increased tension. Manometric reading was not taken owing to the restlessness of the patient. A small quantity of fluid was withdrawn, and it was neither xanthochromic nor turbid. A fibrinous clot did not form on standing. At this stage it was considered that the patient had an intracranial tumour, but its situation was not localized. The boy's condition became rapidly worse and death occurred within three hours of admission.

On examination, the cerebrospinal fluid was clear and contained a normal amount of protein, and 28 cells (mostly lymphocytes) per c.mm. were present. The chloride content was 718 mg. per cent., and the Wassermann reaction was negative.

PATHOLOGICAL FINDINGS

Autopsy (24.5.34).—The body was that of a well-nourished adolescent male.

Brain.—The brain appeared large and oedematous, and there was an excess of clear cerebrospinal fluid at the base. There was no evidence of
meningitis nor were there any haemorrhages. Both lateral ventricles were distended with clear fluid and dilated. Almost filling the third ventricle and attached to the choroid plexus of the velum interpositum was a spherical thin-walled cyst, 1·5 cm. in diameter. The cyst was hanging freely in the third ventricle, but on its superior aspect possessed a vascular point of attachment to the anterior end of the velum interpositum. The third ventricle itself appeared somewhat dilated.

Kidneys.—The right kidney weighed 9 oz., the left 8 oz. Both kidneys showed a polycystic condition. The cysts were not apparent on examining the surface of the kidneys, but on the opening of each organ multiple small cysts were found lying deeply within the cortex, and mainly at or near the junction of cortex with medulla. The largest single cyst was not more than 4 mm. across, and the smallest cysts were of pinhead size. In areas, a group of small cysts formed a honeycomb nodule about 1·5 cm. in diameter. The capsules of the kidneys stripped with ease, and no other changes were seen.

Other organs were carefully searched for cystic disease, but none was found.

Lungs.—An active caseating tuberculous focus was found at the base of the left upper lobe, and a small fibrous nodule was present in the left lower lobe. The left hilar glands were enlarged and showed commencing caseation.

MICROSCOPICAL EXAMINATION

Third Ventricle Cyst.—The contents of the cyst, which, in the fresh specimen, were of a viscid fluid nature and turbid, became brittle with fixation. Under the microscope, the cyst was seen to be filled with a homogeneous eosinophilic material. A few cuboidal epithelial cells and an occasional leucocyte were scattered throughout this hyaloid matrix.

The cyst wall was constructed of thin connective tissue, on the inner surface of which was a lining of epithelium. At one point on the circumference the connective tissue wall became thicker and highly vascular and merged insensibly with the vascular stroma of the choroid plexus (fig. 1). At this point the outer wall of the cyst was in relation to choroid plexus, but elsewhere on the outer surface—that bathed by the fluid in the third ventricle—there was an incomplete much-flattened epithelium, the nature of which it was impossible to determine. The lining epithelium of the cyst consisted of cuboidal cells with round dark nuclei identical with choroid plexus epithelium (fig. 2). These cells were arranged in a single layer round most of the circumference, but at the point of attachment to the choroid plexus the lining epithelium of the cyst became four to six layers deep, although neither invasion of the stroma nor papillomatous formation occurred. These cells did not appear to possess cilia.

Sections were stained by Cajal’s gold-sublimate method, and the connective tissue of the cyst wall was determined to contain a few well-developed astrocytes.
FIG. 1.—Wall of cyst of third ventricle at the point of attachment to the choroid plexus. At this site the lining epithelium of the cyst is four to six layers in depth. × 48.

FIG. 2.—Wall of cyst of third ventricle, showing single layer of non-ciliated cuboidal epithelium. × 440.

FIG. 3.—Section of kidney. The cyst here shown is lined by a single regular layer of cells identical with the epithelium of the renal tubules. × 440.
Kidneys.—The cysts in the kidneys presented the varying features frequently met with in polycystic disease, though the small size of the cysts and the lack of general involvement of renal tissue were unusual. Some of the cysts were lined by a single regular layer of short columnar cells indistinguishable from renal tubule epithelium (fig. 3). Other cysts had a lining of shorter, cuboidal cells, and others a simple flattened epithelium, closely applied to an encircling stroma of connective tissue. A considerable formation of fibrous tissue was apparent around the larger cysts.

Lungs and Bronchial Glands.—The presence of active tuberculosis was confirmed.

DISCUSSION

Clinical.—Diagnosis of third ventricle cysts is, as yet, rarely possible during life, though Zimmerman and German localized the lesion in their cases and the nature of the obstruction was revealed at operation. They lay great emphasis on the localizing value of ventriculography in the investigation of the cases. Many of the patients have been young, half being under thirty-five years of age, and the present case is the youngest recorded. A number of patients have died suddenly, some being found dead; others were found unconscious and died within a few hours. These cases are, therefore, of medicolegal as well as pathological interest. Drennan’s cases (1929) were both the subjects of coroners’ inquests.

The symptoms and signs of this condition naturally fall into two groups: (a) generalized increased intracranial tension, and (b) localizing factors. When the patient is comatose it is likely that the features of the first group will be mostly in evidence. Headache, vomiting, mental dulness, apathy, coma and papilledema were the principal manifestations of increased intracranial pressure in this case, and it is doubtful whether the bilateral extensor plantar responses and urinary incontinence were of any localizing value. The onset of acute hydrocephalus in these cases is due to sudden occlusion of the foramina of Munro or the aqueduct of Sylvius by the cyst. It is possible that the slight blow on the head which our patient sustained just prior to the onset of his headache was sufficient to cause a displacement of the cyst and so lead to the development of hydrocephalus. The occlusion is not always permanent; if it is only temporary and intermittent the symptoms will vary accordingly. In a number of cases it is stated that moving the head in certain directions relieved the severe headaches, the relief being often sudden and striking. This phenomenon, though suggestive of a ball-valve action of a cyst in the third ventricle, is not always diagnostic, as it may occur with tumours of the posterior fossa. Attacks of hypersomnia have been described in association with colloid cysts, and attributed to temporary occlusion in the ventricular system, although in view of the proximity of the hypothalamic sleep centre it has been suggested that other factors have to be considered.

Weisenburg (1911) published the first series of third ventricle tumours,
and attempted to define a third ventricle clinical syndrome. Since then attempts have been made to amplify his findings in the light of recent publications of new cases. Fulton and Bailey, in summing up this question, believe that 'there is no syndrome of the third ventricle, per se, but tumours immediately affecting this cavity may cause characteristic symptoms by pressure upon its walls and their contained nuclei and tracts. Among the well-recognized syndromes due to lesions of these structures we may mention: (1) the infundibular syndrome (polyuria, adiposity); (2) the syndrome of the central grey matter around the posterior end of the third ventricle and aqueduct of Sylvius (hypersomnia); (3) the thalamic syndrome (central pain, painful hyperaesthesia); (4) the extrapyramidal syndrome (bradykinesia, rigidity); (5) the decerebrate syndrome (hypertonicity, Magnus-de Kleijn reflexes; (6) the syndrome of Parinaud (paralysis of conjugate vertical movements of the eyeballs); (7) the syndrome of the body of Luys (hemichorea; vide Martin, 1927, Ewald, 1891); (8) the hypopituitary syndrome (infantilism, hypotrichosis, lowered metabolism); (9) the uncinate syndrome (olfactory and gustatory symptoms; vide Herzog, 1929), etc.' A number of these syndromes—often incomplete—have been found in a few cases to be associated with colloid cysts. It has been indicated however that, while there are often striking manifestations when the floor and lateral walls of the third ventricle are involved by tumours, there are practically no signs pointing to a lesion of the upper part or the roof of this cavity. Högner (quoted by Fulton and Bailey) states: 'Tumours extending into the third ventricle from the roof are remarkable for their lack of symptoms.' The majority of the colloid cysts in this region—as in the present case—appear to have their origin in the roof of the third ventricle, and this accounts for the absence of localizing signs (excluding ventriculographic findings) in most of the cases.

Pathological.—Colloid cysts of the third ventricle are rare and their aetiology obscure. Most of the argument as to their pathology has centred round the question whether they are of ependymal or of choroid plexus origin. Drennan (1929) describes two cases with local features almost identical with those of our case, and believes them to be of choroid plexus origin, a view that is supported by the finding, in one of his cases, of a second much smaller cyst attached to the choroid plexus of the right lateral ventricle. Byrom and Russell (1922), in describing one case, believed that the cyst was ependymal on the grounds that the lining epithelium showed occasional tufts of cilia. But Zimmerman and German (1938) point out that the finding of cilia in these cysts is not conclusive evidence that they are of ependymal origin, because in the embryo the cells covering the choroid plexus also bear cilia. Certain of these cysts appear to develop above, and to be covered inferiorly by the velum interpositum (Hall, 1918; Fulton and Bailey, 1929; Byrom and Russell, 1922), but such cysts may have developed in the choroid plexus of the lateral ventricle and slipped through the foramen of Munro to
COLLOID CYST OF THE THIRD VENTRICLE

come to lie in the third ventricle as described by Hall. The finding of a cyst with ciliated epithelium in this position led Sjövall (quoted by Fulton and Bailey) to suggest it arose from the paraphysis, which is a rudimentary structure in man projecting into the anterior part of the third ventricle, and this is a view that cannot at present be disputed.

We are content, however, to consider our cyst as originating in the choroid plexus when we take into account the similarity of its epithelium with that of the choroid plexus and the clear association of the connective tissue of the cyst wall with the vascular stroma of the plexus.

The association of a cyst in this position with polycystic disease of the kidneys is of interest as it substantiates the impression, generally existing, that the former condition is 'congenital'; that it is, in other words, a local manifestation of an aberrant developmental process whereby certain glandular or epithelial-lined spaces are snared off, with the occurrence, sooner or later, of cystic dilatation. Kauffmann (1929) supports the view that such a process is responsible for the majority of congenital cystic kidneys. He describes it as 'a failure of certain urinary tubules to become patulous, followed by cyst formation.' It is easy to believe in such an origin when one is presented with a microscopical picture like that shown in fig. 3.

Polycystic kidney is quite often only one of many congenital malformations. A case in point recently came within the practice of this laboratory in which a foetus presented the peculiarities of a polycystic right kidney, micrencephaly, bilateral cleft palate and a bicornuate uterus.

SUMMARY

(1) A case of colloid cyst of the third ventricle is described and the symptomatology discussed.
(2) The pathology of this condition and its relation to cystic disease of the kidney, which was also present, are considered.
(3) It is believed that the majority of these third ventricle cysts are congenital and due to a developmental malformation of the choroid plexus.

We are indebted to Dr. T. B. Davie for assistance in the preparation of this paper, and for allowing us to quote his case of multiple congenital abnormalities.

REFERENCES

BATTEN, F. E., and COLLIER, J. S., Brain, 1899, 22, 473.
BYROM, F. B., and RUSSELL, D. S., Lancet, 1932, 2, 278.
HALL, A. J., Lancet, 1913, 1, 89.
KAUFFMAN, E., Pathology, 1929, 1385.
WEISENBURG, T. H., Brain, 1911, 33, 296.