A CASE OF PINEAL CYST

BY

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[WITH PHOTOGRAVURE PLATE]

Neither Cushing's (1932) collection of over 2,000 verified intracranial tumours, nor that reported by Walshe (1931), contained a case of a simple pineal cyst. Prof. D. Russell, in a personal communication stated that she had occasionally seen a small pineal cyst at necropsy, but that this had been an incidental finding and had not caused symptoms. In our own case the cyst was large and caused signs and symptoms of an intracranial tumour.

Clinical Details

The patient, a female aged 21 years, had been well until August, 1946, when she began to suffer from frontal headaches. One day during the same month she was suddenly seized by a feeling of numbness and coldness in the right side of the face, neck, and the right upper limb. When the arm was pinched she felt no pain. The right arm was weak and shook when she attempted to grasp anything. A few minutes later a similar condition developed in the left arm ; normal sensation and power returned within about half an hour and she was left with severe frontal headache and nausea. She stayed in bed all that day and returned to duty on the following morning. During the next four weeks the frontal headache increased in severity and frequency. During September she had another attack, very similar to the first, but this time it lasted three to four hours and was followed by vomiting. From then on attacks of " pins and needles " and numbness in the right arm lasting a few minutes became frequent. A few days before her admission to this hospital on Oct. 18, 1946, diplopia and mistiness of vision had developed, and she now complained of almost continuous severe headache with frequent vomiting.

Examination revealed early papilloedema, bilateral external rectus palsies, and diminished perception of pin-prick and light touch in the right trigeminal area. She could not maintain posture with the right arm as well as with the left. Recognition of pin-prick, light touch, and vibration was impaired in the right upper limb and, to a lesser extent, in the left lower limb. Position sense and two-point discrimination were not affected. Power and tone were equal and normal in all limbs. *X*-ray examination showed no abnormal calcification. Ventriculography was performed on Oct. 21. The lateral ventricles were moderately dilated, the left being slightly larger than the right. There was a rounded filling defect of the posterior part of the third ventricle, and neither the aqueduct nor the fourth ventricle could be visualized.

An operation under general anaesthesia was performed on the same day. A right occipito-parietal bone flap was raised. The intracranial tension remained high in spite of ventricular punctures, and in order to reach the posterior portion of the third ventricle the occipital lobe was removed ; after a little retraction of the parietal lobe a rounded tumour came into view. It lay just beneath the splenium of the corpus callosum and its lower portion was hidden by the tentorium. Dissection of the tumour was not difficult ; the capsule was tough and well defined, and no vessels were seen entering it. Until the final removal the patient's general condition remained good, but as the tumour came away her breathing stopped, and although it was resumed after a brief period of apnoea she died as the wound was being closed.

The Cyst

The cyst, preserved in 10% formol saline, was grey, oval, and was 3.2 cm. long by 2 cm. wide. It had been opened before arrival in the laboratory and was empty when received. It had contained a light-yellow fluid. The wall, which varied from 3 to 5 mm. in thickness, was apparently in two layers ; externally there was a paper-thin and slightly granular layer of greyish tissue, whilst internally and forming most of the wall was a soft but firm yellowish-brown material.

Histologically (see Plate) the wall was really formed of three layers. Externally was a very thin network of glial fibres (Fig. 1, A) covering a middle cellular layer (Fig. 1, B ; Fig. 2), which varied in thickness and was interrupted in places. In general this layer was 10 to 30 cells in depth, but in some areas was up to 100 cells thick. The cells, which were moderately closely packed, had round or slightly oval nuclei with fine or coarse chromatin stippling and thickened nuclear membranes. The cytoplasm was abundant but the cell outlines were indistinct. Some hyaline, concentrically striated, irregularly round bodies (corpora arenacea) were present. Internally was a thicker continuous layer of dense gliotic tissue (Fig. 1, C) containing numerous brown pigment granules mostly within macrophage cells. These granules and some areas of gliotic tissue gave a strong Prussian-blue reaction for iron.

Post-mortem Examination

A necropsy was performed nineteen hours after death. The summarized findings are as follows:

The *brain* weighed 3 lb. (1.36 kg.). Recent right occipital lobectomy. Subarachnoid haemorrhage on the ventral surface of the brain stem. Blood-clots present in the anterior horns of both lateral ventricles and in the third ventricle. The Sylvian aqueduct was patent, the fourth ventricle natural, and no hydrocephalus was noted. The tectum of the midbrain was distorted: on the right side it was compressed, and the left dorsal surface, which projected posteriorly more than the right, was irregular. The superior and inferior colliculi were indistinct. Two small recent haemorrhages were seen in the substance of the pons and midbrain. No pineal gland visible. Histology of the midbrain showed a loss of ganglion cells and a partial demyelination in the region of the colliculi in addition to the distortion. The periaqueductal grey matter was reduced in thickness and cell density but chromatolysis was not observed. Both *lungs* were partially collapsed. The *liver* weighed 2 lb. 3 oz. (995 g.), and was darker and firmer than usual. Histologically some excess of fibrous tissue was seen around Glisson's capsules, possibly an early periportal cirrhosis. The *uterus* was pregnant and enlarged to 1 in. (2.5 cm.) above the symphysis pubis ; it contained a normal male foetus 16 to 18 weeks old. Large corpus luteum in right ovary, secondary areolae of the nipples, mammae firm and containing hypertrophic glands.

Discussion

The symptoms and signs were due to pressure of the cyst on the tectum of the midbrain in combination with internal hydrocephalus. This latter was probably intermittent and was not apparent at necropsy. Prof. D. Russell, who examined sections of the cyst, suggested that it was of pineal origin but of unusually large size. The presence of corpora arenacea, the character of the epithelial cells in the middle layer of the cyst wall, the situation of the cyst, and the absence of the pineal gland at necropsy support this opinion.

It is difficult to say how the cyst arose, but it is likely that intrapineal haemorrhage was responsible, if not for the origin of the cyst, at least for its unusual size, and that the gliosis was secondary to the haemorrhage. As the cyst enlarged, the pineal epithelium became thinned out in a layer surrounding the internal haemorrhage and gliosis. The cyst first caused signs of increased intracranial pressure when the patient was six to eight weeks pregnant. The association between the two conditions—a large pineal cyst and pregnancy—may be no more than a coincidence, but it is interesting at least to speculate whether the pregnancy might have caused a change in a pineal cyst present before, but too small to produce symptoms.

Summary

A case of an unusually large pineal cyst in a young woman is described.

The cyst was large enough to produce local midbrain pressure symptoms and internal hydrocephalus, probably intermittent.

Details of the histology are given.

Our thanks are due to our colleagues, particularly to Major R. C. Connoliy, R.A.M.C., to Prof. Dorothy Russell for giving us her opinion on the sections, and to Dr. P. Daniel for taking the photomicrographs. We are grateful to our commanding officer, Col. G. D. Gripper, for permission to publish this case.

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AMYOPLASIA CONGENITA ASSOCIATED WITH HYPEROSTOSIS FRONTALIS INTERNA

BY

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[WITH PHOTOGRAVURE PLATE]

The absence of any reference in the literature to the combination of amyoplasia congenita and hyperostosis frontalis interna and the striking clinical picture resulting from this compound syndrome prompt me to record the following case.

Case Report

The patient was a woman aged 61 years. Since birth her lower limbs had been abducted at the hips and flexed at the hips and knees, her arms had been extended at the elbows and flexed at the wrists, and movements at all the joints had been severely limited. She learnt to shuffle round the floor, to feed herself, write, sew, knit, and do other forms of handicraft. She went to school from the age of 7½ to 9 years, was fond of reading, and was as apt as children of her own age. She was of a cheerful disposition, being despondent only when afflicted by pain. She had had headaches behind the eyes and over the frontal region as long as she could remember, the more severe attacks being accompanied by nausea and vomiting, but had never complained of her sight or of seeing double. At the age of 9 she had measles. Menstruation started when she was 11, and, although at times periods were regular every 28 days, she was subject to long spells of amenorrhoea even up to four years. She ceased to menstruate at the age of 54. When she was 19 she gained weight rapidly, and soon attained a weight of 11 stone (70 kg.). At the same time she developed in the beard area and on the upper lip a growth of hair which became sufficiently noticeable to require the use of a razor or depilatories.

At the age of 41 she began to get severe attacks of colicky pain in the upper part of the right side of the abdomen, radiating round to the right scapula. A diagnosis of gall-stone colic was made. Eighteen months later she was found to have a duodenal ulcer. This required two years' medical treatment. When 50 years old her gall-bladder was drained but not removed. Through the operation scar there developed a large incisional hernia, which has been a constant source of pain and distress.

At the age of 52 she complained of excessive thirst; sugar was found in her urine. She was given 15 units of insulin morning and evening, without any special diet, for four months. In 1941 she was admitted to Southmead Hospital, Bristol, on account of an abscess in the pubic region. Three years later she was again admitted with multiple boils and an anal fistula, for which she was operated on. Sugar was again found in the urine, and she was discharged on 40 units of insulin a day but no dietary restrictions; this did not render her sugar-free. She was re-admitted to Southmead Hospital on June 6, 1946, because of recurrent attacks of severe pain in the incisional hernia associated with vomiting. There was heavy glycosuria but no ketosis. In the surgical ward, in an attempt to control the glycosuria, the insulin was increased to 40 units four-hourly, but in spite of this each specimen of urine was loaded with sugar. She never had an overdose. So far as can be ascertained from the notes she has never had ketonuria.

Family History.—Her father died at the age of 80. Her mother, who died at the age of 84, had nine pregnancies. The first was a boy, who died at 14 months after an accident; the second child was a girl, who is still alive, and whose only complaint has been cholecystitis. The next in the family was the patient, who was born when her mother was 36. There is no knowledge of any complications during the pregnancy or confinement. All the remaining pregnancies ended in miscarriages at the fourth month. There was no consanguinity either in the parents or in the grandparents.

Physical Examination.—The gross obesity coupled with the limb deformities made a striking picture (Plate, Fig. 1). Sitting height 32 inches (81 cm.), weight 11 st. 9 lb. (74 kg.). She had a large fat ruddy face with hair on the chin and upper lip. The lobes of both ears were webbed. The neck was short, and the neck and body were grossly obese, which contrasted with the thin upper and lower limbs. Abdominal fat sagged down and partially covered the drawn-up legs. The breasts were normally developed, and the hair on the head and body was of normal female distribution. The skin was greasy and subject to acne, but there were no striae or abnormal pigmentation.

Passive and active movements of the joints were approximately equal. Abduction of the shoulder-joints was limited to 15°. The arms were extended at the elbows and the forearms pronated. Movements of the elbow-joints were limited to 5°. The wrists were fixed in flexion and ulnar deviation. The finger-joints were flexed. She was able to flex fingers and thumbs to about 60°, but had practically no power of extension in the fingers. She was able to oppose her thumbs to any of her fingers, and thus grip an object between the fingers and thumbs. There was webbing of both axillary folds and of the first and second fingers of both hands. There were flexion and abduction at the hip-joints, flexion at the knee-joints, and bilateral equino-varus. She could flex the right hip-joint about 40°, and had moderately good power of abduction and adduction of the right femur. Movement of the left hip was more limited than that of the right. Movement was extremely limited in the knees and was absent in the ankle-joints. There was limited flexion of the toes. She had slight scoliosis, and mobility of the back was very limited.

The pupillary responses to convergence and accommodation were normal. Eye movements were full and the media and fundi were normal. Peripheral and central fields of both eyes were full to 3/330 and 3/1,000 mm. white object, respectively. Her visual acuity in the right eye was 6/9 and in the left 6/6 without glasses, and 6/6 in both eyes with correction. She had complete upper and lower dentures. There was no thyroid enlargement and no abnormal pulsation in the neck. The pulse rate was 70 per minute and the blood pressure 170/90 mm. Hg. The heart and lungs were normal. There was a large incisional hernia on the right side, painful on palpation. The liver and spleen were not felt and there was no glandular enlargement. Nothing abnormal was found on examination of the central nervous system.

Investigations.—Blood: Hb, 116% (Haldane); R.B.C., 5,720,000; W.B.C., 8,200 (neutrophils 62%, lymphocytes 31%, eosinophils 2%, monocytes 5%). Urine: specific gravity, 1027; acid; no albumin; sugar, 260 mg. per 100 ml.; no ketone bodies; urobilinogen not increased; bilirubin absent; deposit not remarkable. Blood calcium, 10.4 mg. per 100 ml.; blood phosphorus, 4.3 mg. per 100 ml.; serum phosphatase, 9 units; 24-hourly urinary excretion of calcium, 320 mg. Blood Wassermann and Kahn reactions negative. Both the glucose-tolerance and the insulin-tolerance estimations were performed at a time when the patient had been receiving a normal mixed diet. There was an insulin-resistant diabetes, as the following figures show: The blood cholesterol was 185 mg. per 100 ml. It was estimated on many ocasions, but was not found to vary appreciably with the blood-sugar levels. The 24-hourly 17-ketosteroid excretion was determined on two occasions, and gave figures of 2.3 and 4.1 mg.

X-ray Examination.—Heart and lungs were normal. A congenital deformity of the first and second ribs on the right and a short first rib on the left were present. There were a bony bridging between the vertebrae, slight scoliosis, and fixation of

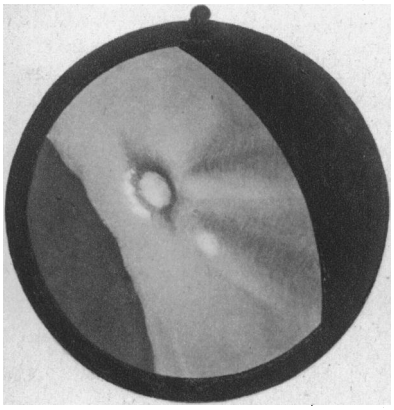

FIG. 1.—Small acute ulcer, healed five days later.

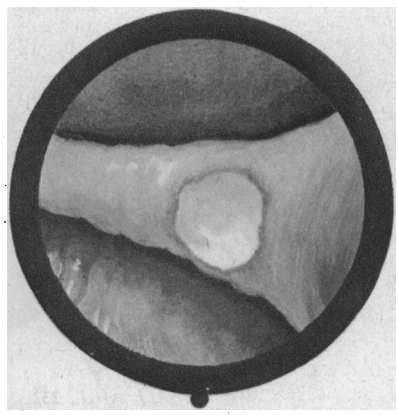

FIG. 2.—Acute ulcer on a mid-gastric constriction ring.

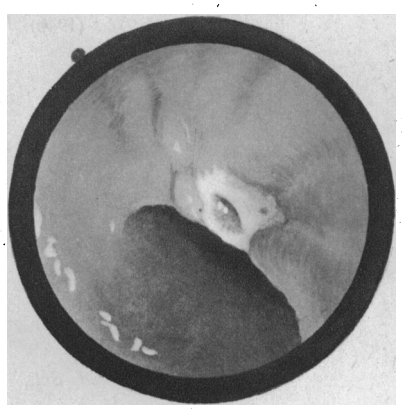

FIG. 3.—Subacute ulcer.

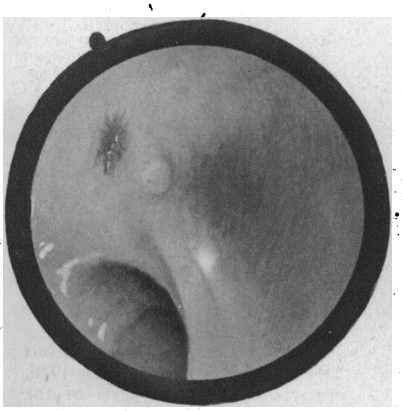

FIG. 4.—Same ulcer as Fig.3, healed after two weeks.

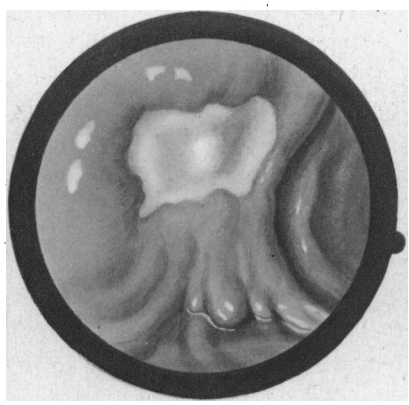

FIG. 5.—Subacute ulcer.

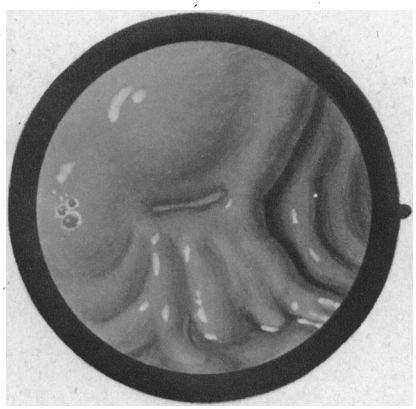

FIG. 6.—Same ulcer as Fig.5, almost healed after two weeks.

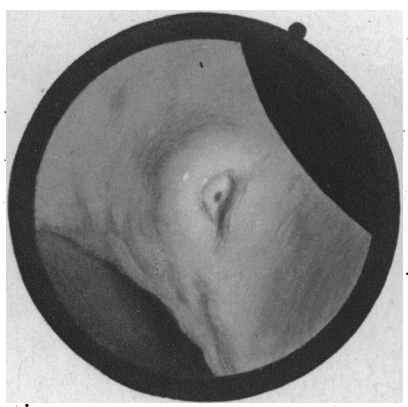

FIG. 7.—Subacute ulcer, posterior wall.

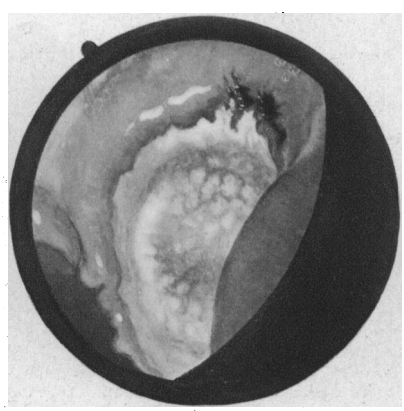

FIG. 8.—Large ulcer, posterior wall.

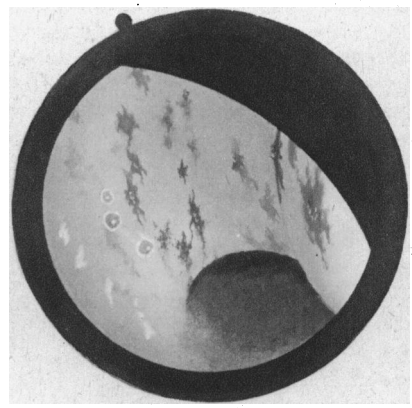

FIG. 9.—Multiple intramucosal haemorrhages.

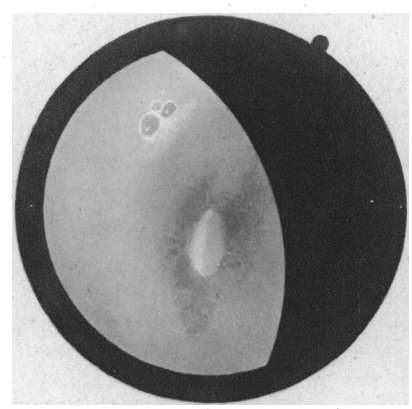

FIG. 10.—Haemorrhages disappeared in one week, leaving small acute ulcer.

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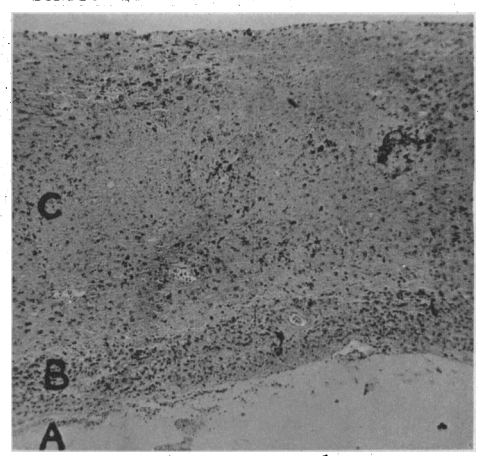

FIG. 1.—Thin glial layer externally (A), middle epithelial layer of pineal parenchyma (B), and internal pigment-containing gliotic layer (C).

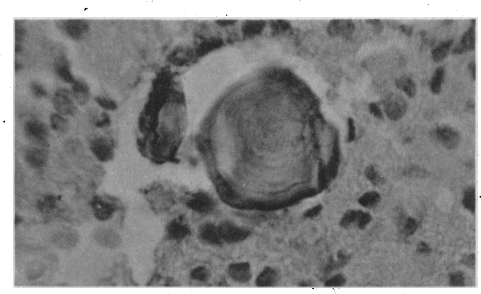

FIG. 2.—High-power view of B, showing epithelial cells and one corpus arenaceum.