A HUGE OSTEOMA IN THE ANTERIOR CRANIAL FOSSA

BY

VI. HUDOLIN, D. RIESSNER, S. KADRKA, and M. KNEŽEVIĆ

From Neurološko-Psijihijatrijski Odjel, Zagreb

Osteomata with intracranial complications are rare, so that even today they attract interest (Campbell and Gottschalk, 1938; Hariga, 1960), especially when they are not arising from the paranasal sinuses, where they usually originate.

Recently we had occasion to observe a patient with an osteoma of unusual size situated in the anterior cranial fossa. The clinical picture was that of a case of frontal lobe syndrome with personality changes and symptomatic epilepsy. We are describing the case because of the unusual site of the osteoma and also as a contribution to the diagnosis of the psychiatric syndrome in lesions of the frontal lobe.

Osteomata in the intracranial cavity must be distinguished from secondary bone formations and from the so-called osteoplastic meningioma (Cushing, 1937). According to the medical literature they are very rare, and are usually found either as small bony formations of the cranium, or as exostoses in the region of the anterior clinoid processes, the lesser wings of the sphenoid bone, or pyramidal portion of the temporal bone (*porus acusticus internus*) (Lindgren, 1954; Ritvo, 1949; Coley, 1949). Osteomata in these sites show very little tendency to grow. More frequently osteomata are found in the paranasal sinuses or arising from the orbital roof, and these orbitoethmoidal forms can lead to complications following their penetration into the intracranial cavity. Frontal lobe complications in the cases hitherto described arose after penetration of the accompanying mucocoele into the intracranial cavity then into the brain substance and finally into the ventricles (Cushing, 1927; Campbell and Gottschalk, 1938; Hariga, 1960). Spontaneous pneumocephalus and meningitis are the complications usually described. Peters (1951) mentions that osteomata of the cranium are relatively rare and gives Courville's classification, namely, 1, circumscribed osteoma of the cranial vault (single or multiple); 2, diffuse osteoma (usually arising from the great wing of the sphenoid or from the temporal bone); 3, osteoma

of the orbito-ethmoidal region; and 4, osteochondroma arising from the base of the skull.

Cushing (quoted by Zülch and Christensen, 1956) had in his series of 2,023 tumours only 14 osteomata. Zülch and Christensen (1956) in their own 4,000 cases of cerebral tumour mention 16 osteomata, while Olivecrona (quoted by Zülch and Christensen, 1956) had no cases of osteoma in his series of 5,250 cases.

The frontal lobe syndrome presents a psychiatric clinical picture which although well recognized cannot be accurately localized or aetiologically determined by clinical methods. When this syndrome has existed for some considerable time, and when there is also severe damage to the frontal lobe, the question then arises whether restoration of mental faculties is in any degree possible, especially when the patient has only psychiatric symptoms which do not allow of accurate objective measurement because the personality changes resulting from frontal lobe lesions cannot be satisfactorily measured by the usual psychological tests. Frontal lobe tumours are usually found in patients in mental hospitals since they very often present only psychiatric symptoms uncharacteristic of any definite clinical picture.

Case Report

A peasant woman aged 57, living in the neighbourhood of Zagreb, was admitted to the Neuropsychiatric Department as an emergency case. At the time of admission no details of her illness could be obtained because of her severe mental derangement and poor physical condition. According to the statement of her husband there was no family history of neuropsychiatric relevance. Since 1937 she had suffered from epileptic fits. She had been prone to drink heavily since early youth, and especially since the end of the Second World War. She had already received hospital treatment in 1954 having been treated as a chronic alcoholic with antabus. Shortly after hospital treatment she suffered from an extremely severe attack of epilepsy, being unconscious for one hour followed by a disturbed mental state for several days. Visual hallucinations were

present. She was again admitted to a mental hospital and discharged after 18 days as a case of chronic alcoholism with symptomatic epilepsy; she was treated with anticonvulsant drugs.

Her husband remarked that after discharge from hospital on this second occasion she was 'a changed person'. She was restless, forgetful, and careless in her behaviour. Her condition slowly deteriorated, her epileptic attacks occurred once every two or three months, but she did not take anticonvulsants regularly. She continued to drink heavily.

Three months before her admission to our hospital she began to complain of 'terrible headaches' and after a particularly severe headache she lapsed into coma and was admitted to the provincial hospital where she was diagnosed as a case of apoplexy. She was comatose for the first three days after admission. At the end of 12 days she was sufficiently recovered to be allowed home, at the request of her family and against medical advice and before neuropsychiatric investigations had been done. From that time her mental state rapidly deteriorated. She was confused, she spoke at times about herself disconnectedly, saying that it would be better to keep quiet but that she had to speak, etc. At this time she was eating little and drinking heavily and she lost a great deal of weight. Finally, she was admitted to our hospital. On admission she was incontinent of urine and faeces. She responded to questioning, but quite irrationally. She thought that she was in her native village school. She would not change her opinion about this in spite of it being pointed out to her that she was surrounded by hospital beds and not by school desks. She could not grasp who the people around her were. She showed no spontaneous interest in her surroundings, but smiled constantly in a good humoured manner. She offered no resistance to medical or nursing care and was indifferent to injections. She gave the impression that she had no comprehension of what was going on around her. Occasionally she would join in the conversation of the other patients with irrelevant remarks. Among a great many incorrect statements about herself she would make an occasional correct one. She could understand and carry out simple instructions. She would sometimes reiterate a chance word heard from the conversation of others. She had no complaints of any pain.

Clinical Findings.—She was able to walk by herself, but her gait was unsteady with some dystasia. When brought food, she was able to eat unaided. There was no tenderness of the skull on palpation or percussion. No signs of meningeal irritation were present.

The pupils were slightly contracted, equal in size, not distorted; the reactions to light and accommodation were a little sluggish. It was impossible to ascertain the presence or absence of anosmia because of her mental state.

Sensation and the rest of her neurological state could not be estimated accurately but were apparently normal. Her psychiatric state suggested that she was a psycho-organic case with frontal lobe syndrome.

Of the investigations carried out, we cite only the abnormal: The E.S.R. was 20/47 (Westergren), and the

number of cells in the cerebrospinal fluid was increased (34/3). The aorta showed some degree of atheroma in a chest radiograph. The ocular fundi at first showed no abnormality but just before operation a papilloedema of 1D had developed on the right side.

X-ray Findings in the Skull.—Antero-posterior and lateral views of the skull and frontal and sagittal tomography of the anterior cranial fossa (Figs. 1, 2, 3) showed that the skull was normal in shape and size. An opaque shadow approximately 7 cm. in diameter was seen lying on the lamina cribrosa equally on either side of the midline. The density of the shadow suggested a homogeneous calcified circular growth with clearly defined and irregular outlines. The lesion extended from the frontal bone to the sella turcica. There was a projection from the edge of the growth on the right side towards the lateral angle of the right orbit with ill-defined edges.

The internal lamina of the vault was thickened, especially in the frontal area. The caudal part of the posterior wall of the frontal sinus and part of the roof of the right ethmoid sinus were narrowed. The lesser wings of the sphenoid bone were thickened. The sella turcica appeared normal and the dorsum sellae was rarefied.

Pneumoencephalography with 40 ml. of air was done, and showed that the frontal horns of the lateral ventricles were shortened. The central parts of both lateral ventricles were widened and, together with the temporal horns and the narrowed basal cisternae, were displaced backwards. These findings suggested that there was a single multicentric osteomateous tumour, the size of an orange, situated on the base of the anterior cranial fossa lying centrally. The growth had produced backward dislocation of the atrophied frontal lobes, ventricular system, and basal cisternae. There was also erosion of the lamina cribrosa on the right side and of the caudal part of the posterior wall of the frontal sinus, and reactive internal hyperostosis, especially in the frontal bone.

Since the lesion shown on the picture suggested an expansive growth of homogeneous structure the differential diagnosis lay between a calcified olfactory meningioma and an osteoma in the anterior cranial fossa. On the basis of the history and the clinical and radiological findings, we diagnosed the case as a psychiatric frontal lobe syndrome caused by an expansive osteomateous growth in the anterior cranial fossa. It was decided that neurosurgical treatment should be undertaken, especially as signs of raised intracranial pressure were now appearing (papilloedema).

A wide osteoplastic craniotomy was performed by Tönis method. After section of the falx, free access to the base of the anterior cranial fossa was obtained. The dura mater was not tense. After the dura was opened a thick gelatinous mass, which enveloped the tumour to a thickness of 1 cm., began to exude through the opening. Similar gelatinous masses are found in all instances producing stagnation of the cerebrospinal fluid and they contained a great deal of cholesterol. The tumour, which appeared as a white chalky mass, was found to be lying on a broad base on the lamina cribrosa (Fig. 4). It was easy to separate the tumour from the

FIG. 1

FIG. 2

FIG. 5

FIG. 3

FIG. 4

FIG. 1.—Pneumoencephalogram, lateral view, showing the tumour in the anterior cranial fossa and backward displacement of the ventricular system.

FIG. 2.—Pneumoencephalogram, antero-posterior view, showing the tumour centrally placed.

FIG. 3.—Tomogram, lateral view, showing the relation of the central part of the tumour to the floor of the anterior cranial fossa. There is no connexion with the paranasal sinuses.

FIG. 4.—Tumour after removal.

FIG. 5.—Tumour consisting of thickened bony trabeculae and marrow spaces. $\times 100$.

base of the skull with a chisel. After the removal of the tumour the gelatinous mass was evacuated with a sucker and a large cavity surrounded by the atrophied frontal lobes remained. In the bottom of it the medial formation of the brain was seen. The fact that there were no serious post-operative midbrain reactions was mainly due to the presence of the gelatinous mass which gave relatively easy access to the tumour, and consequently irritation of the midbrain was avoided. The post-operative course was normal.

Pathological Findings.—The tumour was of even consistency on section, and after decalcification it was found to consist of spongy bone tissue containing a small number of normal bone cells and with a normal amount of calcium in the trabeculae. The marrow spaces were narrow without haematopoietic tissue and with fibrous connective tissue and wide blood capillaries (Fig. 5). The tumour compressed the brain substance, producing local necrosis with destruction of the granular cells and an accumulation of crystalline cholesterol. The histological diagnosis was osteoma spongiosum.

After operation the patient's condition quickly improved. She was no longer incontinent, she was well orientated, replied rationally to questions, carried out directions, ate unaided, and even assisted the other patients. She recognized the nurses and recollected that she had seen them before operation, but she could not remember their names. She knew the principal saints days, and remembered all the family facts correctly, and that she had completed the fourth year at the primary school. She was able to do simple arithmetical sums, but quickly became confused if asked to do any more complicated calculation. She read and wrote as well as one could expect for the amount of schooling she had received. She no longer had any delusions or hallucinations. She remembered well events of the distant past, but for recent events of the last few months she had only slight recollection. She had insight into her present condition, she was grateful for her treatment, but there still remained a certain superficiality and lack of responsibility in her behaviour.

Testing olfactory sense, which was impossible before operation, now showed complete anosmia.

After the operation psychological testing revealed no major intellectual deficiency.

Antero-posterior and lateral radiographs taken two-and-a-half months after operation showed that the shadow in the anterior cranial fossa of the previous radiographs was no longer visible. There was a light hyperostosis of the right lesser wing of the sphenoid, and shadows showing the site of the craniotomy.

She was discharged well, and advised to continue to take anticonvulsive drugs although during her stay in hospital she had had no epileptic fits.

Conclusion

This patient had an extremely large osteoma which had slowly developed and escaped diagnosis. From the pathological point of view it was interesting as a rare example of a case where it was possible to decide definitely that the growth was a true osteoma.

The problem of long-standing damage to the cerebral tissue merits special attention. In our case the huge tumour produced almost complete atrophy of the frontal lobes with resultant gross impairment of intellect accompanied by euphoria, personality changes, and alcoholism, all of which had lasted many years. Very soon after operative treatment the clinical picture showed an extraordinary degree of improvement both physical and mental. This case proves the enormous capacity for restoration of function after lesions of the frontal lobes have been removed.

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