OSTEOGENETIC DURAL ENDOTHELIOMA: THE TRUE NATURE OF HEMICRANIOSIS.

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IN 1811 Everard Home read a paper with the title 'Cases and observations which shew that inflammation is sometimes communicated from the dura mater to the pericranium.' One of the eight cases reported came to autopsy. The only abnormal condition found in this examination was a tumour which made a projection of half an inch on the right parietal bone. On the under-surface of the bone was another similar tumour; both were 'of a fibrous bony structure. The cranium was sound between them, only unusually vascular.' An engraving from a transection of the bone bears out the description and rather strikingly resembles some of the specimens collected for this paper.

On the suggestion of Mr. Percy Sargent,* the records of the National Hospital, Queen Square, London, were searched, in the spring of 1921, for cases of brain tumour associated with cranial hyperostosis. Four hundred and twenty case-histories were reviewed, in each of which, after microscopical examination, a diagnosis of brain tumour had been made. In ten of these cases a hard lump had been described on the cranium. In each instance the neoplasm was beneath this prominence, and, unexpectedly, all of the cases presented the same histological picture. They fell into the class which is commonly called dural endothelioma. The overlying cranial enlargements proved to be, not simple hyperostoses, but part of the neoplastic process, as will be described below.

There have been isolated reports of examples of the same type of tumour as those reported here, and several cases have appeared in French literature, called hemicranioses. The true histological structure of these cranial prominences has been largely overlooked.

Brissaud and Lereboullet³ described two cases presenting a frontal enlargement limited chiefly, though not entirely, to one side of the head. It was believed, probably because of its hardness, that this cranial boss was a simple exostosis; and as the enlargement to a great extent was limited to the distribution of the upper division of one trigeminal nerve,

* I wish to take this opportunity of expressing my thanks to Mr. Sargent and to the other members of the honorary staff of the National Hospital for permission to refer to cases that have been under their care.

the condition was thought to be the counterpart of the so-called facial hemiatrophy of Romberg. The authors therefore chose to apply the name 'hemicraniosis' to the condition. Parhon and Goldstein,¹⁰ Parhon and Nadjede¹¹ and others have added cases under this name. The striking similarity to certain cases subsequently reported and to cases in this communication is evident. One may assume that, had the bone been examined, it would have been found infiltrated with neoplasm similar to that lying beneath.

Bruns,⁴ Barling and Leith,² Spiller,¹⁴ Tattersall,¹⁵ and Ashurst¹ have added similar cases with the diagnosis of endothelioma. It is

FIG. 1.—Islands of bone forming in dural endothelioma.

evident that the cranial prominence in each instance is not due to a trophic change in the distribution of the trigeminal nerve. Also, the condition is frequently not unilateral. Hemicraniosis is therefore an evident misnomer.

Cushing⁷ has recently made a masterly survey of the subject of dural endothelioma, drawing conclusions from a large series of operative cases, and he described⁶ a typical example of infiltration of the overlying bone by such a tumour causing a hyperostosis. He states that of eighty endotheliomas twenty were accompanied by "an overlying hyperostosis cranii," although no statement is made as to how frequently the bone was studied with the microscope.

The term *dural endothelioma* is employed in this paper simply because

of the general usage of the term. There is no evidence that the tumours arise from endothelium; they are probably derived from the arachnoid, as suggested first by Schmidt,¹³ also by Cushing and Weed,⁵ Weed,¹⁶ and Mallory.⁹ *

The cellular appearance of the intracranial portion of these osteo-genetic endotheliomas, if they may be so called, is much the same as that of the ordinary subdural endothelioma. Grossly they are similar also. They are adherent to the dura and are frequently situated near the falx, most often in frontal or parietal regions, and apparently never

FIG. 2.—Canals of much-eburnated frontal boss filled with endothelioma cells.

below the tentorium. They are circumscribed and do not infiltrate brain tissue, but grow very slowly to a great size.

The cranial part of the growth is unique. In the cases of shorter duration the overlying bone, though hard on external palpation, appears to be more spongy than normal cranium. Endothelioma cells in strands and columns fill the cancellous spaces and Haversian canals (*Fig. 1*). In the cases of longer duration the prominence on the skull has usually been very hard and large. Tumour cells may still be found in the bony canals (*Fig. 2*), and a pad of growth is present on the summit

* Cushing has suggested the use of the term *meningioma* "as a compromise." This is a step in the right direction, but the term is so broad as to include some tumours which do not belong to the histologically clear-cut group which he has helped us to recognize under the name of dural endothelioma.

between bone and scalp (*Fig. 3*). The overlying temporal muscle and scalp are sometimes infiltrated, although the intracranial portion of the tumour may be well encapsulated. There are no metastases.

Instead of destroying the bone which lies in its path, as is usually the case with invading neoplasms, it grows into the cranium and stimulates further bone formation. As Dr. W. C. Clarke aptly expressed it, after examining some of the author's microscopical preparations, the endothelioma in its growth seems to respect the normal process of bone formation. In parts where osteogenesis is most rapid, the tumour cells are separated from new bone by quite a definite connective tissue layer

FIG. 3.—Depression on the outer surface of cranial boss. Same case as Fig. 2.

(*Fig. 4*), whose structure resembles that of the septa which pass through cellular portions of the tumour (*Fig. 5*). The cells of this connective tissue layer resemble the osteoblasts seen in ordinary bone repair. They apparently are responsible for the formation and also, perhaps, for the absorption of bone. In places this layer is extremely thin. The origin of these bone-forming cells may of course be the dura whose fibres are split by the endothelioma cells (see *Fig. 5*), or the osteoblasts already present in bone may have been stimulated to increased activity.

The cases will not be reported in detail here, since this is being done in another communication.* The signs and symptoms, however, with their neurological aspects, will be briefly summarized.

* The cases in detail and their surgical aspects are to appear elsewhere.¹²

The clinical course of the following cases proved to be remarkably consistent. One patient had noticed no lump but observed that his forehead was changing its shape. The other nine patients had all noticed a cranial lump before admission. In four cases this prominence had been of ten years' standing; in four others, two to three years, and had been noticed for one year in one case. In five cases the lump was described as hard, bony, or hornlike before operation, and in the other five it was apparent, from post-operative or post-mortem examination, that the tumour must have been hard on palpation. Two of the three tumours which grew in the temporal fossa were tender on pressure, and one was

FIG. 4.—Dural endothelioma and bone separated by the characteristic layer of connective tissue cells associated with rapid bone formation. Same case as Fig. 1.

quite soft when first noticed. Only one other cranial prominence was described as tender on pressure. Five were insensitive.

The cranial enlargement was observed before the appearance of any symptoms in four cases, and in two of these there was an interval of nine years after the lump was noticed before the appearance of any symptoms. Three patients first noticed the lump about the time the first symptom appeared.

Of the nine cases with adequate clinical histories and microscopic examinations, seven patients complained of headache as their first symptom. These were not usually of the type caused by increased intracranial pressure, but were described as stabbing or neuralgic, and

may be considered referable to the dura; for, at least in four cases, the pain was referred locally to the vicinity of the lump. The following cases may serve as illustrations.

H. P. (case 1) had observed an increasing bony prominence on the left frontal region for ten years. Headache had been complained of only for one year. It was localized to the region of the prominence, and for the last six years had been stabbing in character. (*Figs. 2, 3, and 5* are from this case.)

On the other hand, C. H. (case 5) had had localized pain in the right eye for ten years before admission, while the hard swelling in the tem-

FIG. 5.—Dura, with some of its tissue apparently continuous with the tumour septa.

poral region had been of only two and a half years' duration, with proptosis of the right eye for one year.

J. T. (case 3) had observed a lump on the vertex of the skull for ten years, and for an equal length of time had suffered from epileptoid seizures, these being his first symptom. The history of this case, which is rather incomplete, does not mention headache.

E. D. (case 4) complained of unlocalized "neuralgic pains" in the head for seven years, the cranial boss being of only two years' duration.

In addition to headache the symptoms complained of depended, as one would expect, on the location and size of the intracranial tumour, and were as follows: varying degrees of hemiplegia—three cases; giddiness—three cases; epilepsy, aphasia, diplopia, and paræsthesia—each

two cases; vomiting, amblyopia, mental change, drowsiness, and coma—each one case.

The usual sites of election of osteogenetic endotheliomas may be illustrated by considering those reported here with the eight cases from the literature which are susceptible of analysis. Seven were in the frontal bone, seven in the parietal bone, and four in the temporal bone. A number of them approached and partly crossed the midline. There is no record of such a tumour in the occipital or suboccipital region. The youngest patient being eighteen and the oldest sixty, the average age was thirty-nine. Twenty-nine per cent. of the patients have been female and 61 per cent. male.

If the diagnosis is made sufficiently early, before the invasion of the brain has become extensive, the prognosis in this type of brain tumour is excellent. In this series, of the nine cases with microscopical study, seven were operated upon by Sir Victor Horsley and two by Mr. Sargent. One patient, who had a very large bony tumour with little encroachment on the brain, is alive and well so far as this condition is concerned, sixteen years after operation. She shows no evidence of recurrence. One patient was perfectly well eight months after discharge, but cannot now be located. A third patient was discharged showing slight evidence of a hemiplegia, which was rapidly clearing up. One patient, an energetic clerk in a London bank, is perfectly well now, thirteen years after Sir Victor Horsley removed a tumour, which had infiltrated the right orbit and temporal muscle, and presented a subdural portion weighing 107 gm. A fifth patient is perfectly well and restored to active work as a postman, fourteen months after Mr. Sargent removed an endothelioma infiltrating scalp, skull and parietal lobe. The other four presented large intracranial growths, and died from the immediate effects of the operation, six hours, eighteen hours, twelve days and six weeks after operation.

One of the patients, a registered nurse, out of curiosity concerning a slowly growing lump on her head, consulted Sir William Gowers. She had had neuralgic headaches for several years. Sir Victor Horsley successfully removed the lump and underlying endothelioma. Thanks to their clinical insight, the true nature of the excrescence was recognized, with as yet no evidence of cerebral invasion.

A study of the case histories shows that there was a time when seven of the patients had only the bony swelling associated with local pain. The other two presented a long interval when the bony prominence and epilepsy were the only complaints. If the medical profession were better acquainted with the condition, it should be possible in a large percentage of these cases to advise operation before the onset of cerebral symptoms. The typical slow-growing bony prominence associated

with local pain, often of a stabbing character, should be considered pathognomonic of the condition.

Neoplasms of this type show no tendency to local recurrence after removal, even when scalp and muscle are invaded. They do not form a fungus when incompletely removed. It is possible that after removal of the cranial and intracranial portion of the tumour the scalp invasion does not continue to grow. There is, of course, a considerable immediate operative risk due to the great vascularity of the involved bone. Horsley's method was to cut through normal skull and rapidly remove the whole tumour *en masse*. The operative difficulties are much decreased if the diagnosis be made early.

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