VASCULAR TUMORS OF THE BRAIN AND SPINAL CORD AND THEIR TREATMENT

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Blood vessel tumors of the nervous system have given rise to considerable discussion for many years. Virchow9 was the first to point out that, as a rule, they are of congenital origin.

In 1915, one of us (E.S.8) described a number of these lesions and, in order to make it clear that they are not true tumors, suggested the term intracranial telangiectasia. This term has not been generally accepted, though Cushing and Bailey,2 in a comprehensive monograph on these "tumors" in 1928, divided them into four groups and used this term to describe the first of their groups, which were as follows: (1) Telangiectases; (2) angioma venosum; (3) angioma arteriale; and (4) hemangioblastoma.

That the first three types are not true neoplasms is well recognized by pathologists, and in his recent Textbook of Pathology Robert Moore7 brings this point out clearly. In the opening sentence of the section on tumors of vessels, he says: "Benign tumors of vessels are of two types, hemangioma and lymphangioma. Most examples are not arteriovenous neoplasms but remnants of fetal tissue misplaced or disordered in development. . . ."

He further points out that, of the four types Cushing and Bailey describe, the only one that may be called a true neoplasm is the hemangioblastoma. This is the view that one of us (E.S.) has maintained and taught for many years. Cushing and Bailey make the statement: "From our personal experience, it is evident that surgery, at its present state of development, offers little as a means of controlling one of these lesions in the brain by direct intervention and any attempt at their operative removal is foolhardy."

In 1929 we attempted for the first time to treat one of these lesions by coagulation (Case 27). Because the treatment of these tumors constitutes a problem of its own, it has seemed to us of interest to collect and report our experiences. Before entering into a discussion of this problem, however, we are obliged to state that we have found Cushing and Bailey's classification unsatisfactory, since both clinically and pathologically it is not possible to differentiate the four types they describe. There are imperceptible gradations ranging from telangiectases to venous and arterial angiomata in which one or the other type of vessel predominates. The one type that is a distinct entity is the hemangioblastoma, which is a true tumor. In this paper, however, we are considering only those cases that fall into the first three groups.

Certain men's names are associated with descriptions of these tumors, notably von Hippel,5 Lindau6 and Dandy.3,4 These men have made notable

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contributions which have tended to clarify some phase of the subject. von Hippel in 1904 described an angiomatous condition in the retina; Lindau in 1927 pointed out that these lesions might be multiple and that when they occurred in the eye there were frequently lesions also in the brain. In 1930 one of us pointed out that, since in certain cases of polycythemia choked discs had occurred, the possibility that a tumor might be present should be considered in every instance. Carpenter, Schwartz and Walker\(^1\) in 1943 proved that this was true and demonstrated in two cases of so-called polycythemia vera that the underlying cause actually was an angioma of the cerebellum, and that, when this tumor was removed, the blood picture became normal. We had such a case in 1944 (Case 10). Whether eye-ground changes that are indistinguishable from choked disc may occur in polycythemia and yet not be associated with tumor is as yet an unsettled question.

All the patients in this series have been operated upon by one of us (E.S.) or by Dr. Leonard T. Furlow. In this series there are 28 cases of angioma. Of these, 7 occurred in the spinal cord; the other 21 occurred in the cerebral or cerebellar cortex. All lesions, both cranial and spinal, fall into one of three categories: telangiectasis, angioma venosum, or angioma arteriale. At times it was impossible to determine to which group a case belonged. In the early cases, the symptoms are indistinguishable and in the well developed cases, it is often quite impossible to distinguish between an angioma venosum and an angioma arteriale. The classification made by Cushing and Bailey, therefore, is quite arbitrary, since not infrequently there is a mixture of the two latter types (Case 13). Hence, from a clinical point of view, whether of treatment or prognosis, it is entirely adequate to call these lesions either telangiectases or angiomata.

**DIAGNOSIS**

The diagnosis of these conditions prior to operation is very difficult to make; in fact, in most cases it may be little more than a suspicion. Although we have, on several occasions, suspected such a lesion, an absolutely positive preoperative diagnosis has rarely been possible. Occasionally the diagnosis may be made from the roentgenogram, as in Case 13 (Fig. 5). In this case, many of the larger vessels of the angioma were calcified but, at operation, the picture that presented was very different, for, in addition to the larger vessels seen in the x-ray, there was a mass of capillary vessels (Fig. 6). In the spinal cases, there is usually nothing distinctive. Of course, when a patient with a large skin telangiectasis, as in Case 12, suddenly develops a paraplegia with a level corresponding to the site of the skin lesion, the diagnosis is obvious, or, as in Case 21, a very unusual history may make such a diagnosis probable. In the cranial cases, the history of prolonged Jacksonian convulsions, without pressure symptoms and without the history of trauma, should make one suspect such a lesion. If, in addition, there are lesions in the skin, this becomes more likely. If there is a vascular abnormality of the retina, as in Case 19, an intracranial blood vessel lesion is probable.