A brother and sister came under our observation with expanding intracranial lesions of similar nature. We thought it would be worth while to report these cases, especially in view of the conclusions drawn by Munslow and Hill[1] in their paper on familial gliomas: namely, that one way to cast some light on this pathogenetic problem, still quite obscure, would be to report all cases of such occurrences, in order also to gain a better perspective on the real incidence of these familial lesions.

It is not entirely uncommon that members of the same family may be affected by similar intracranial expansive processes, particularly neurofibromas, as part of von Recklinghausen’s disease; numerous reports can be found in the literature as proof that these neoplasms have familial and hereditary tendencies.[2,10,14]

Cerebral gliomas have also been reported as occurring in members of certain families.[3,4,11,12,17,19] Cases of cerebellar medulloblastomas affecting twins were reported by Leavitt.[13] Norlén[16] and Adams[1] published cases of vascular intracranial tumors (angioblastomas) showing familial tendencies. Craig[4] reported the cases of two sisters who at approximately the same age presented similar syndromes: at surgery he found a craniohypophygioma in one and a glioma of the optic chiasm in the other.

As for the meningiomas, only the angioblastic variety is said to have certain familial characteristics. These angioblastic meningiomas, however, were shown to have peculiar histologic similarities to the cerebellar angioblastomas of Lindau’s disease,[9] which is notably heredo-familial in nature.

Wolf and Cowen[20] reported cases of Lindau’s disease with supratentorial angioblastomas, probably quite similar to the parasagittal angioblastic meningioma which Patterson and Anderson[18] found in one of two brothers whose cases they reported.

CASE REPORTS

Case 1. No. 402/54. A.M., a 39-year-old female, was admitted on Oct. 27, 1954. One and a half years previously she had a first episode of sudden loss of consciousness. A similar episode occurred 6 months later. At that time she started complaining of frontal headaches and dizzy spells. One month before admission she noticed that her vision was beginning to fail. She consulted an ophthalmologist who referred her to our service.

Examination. The patient showed marked pyramidal signs on her right side, together with a certain suggestion of “Witzelsucht.” There was papilledema of the optic discs; visual acuity was 4/10 in the right eye and 1/30 in the left. The electroencephalogram showed a slow-wave abnormality in the left hemisphere. Left percutaneous carotid angiography showed a moderate flattening of the carotid syphon, with the contrast medium homogeneously dispersed in two round areas, in the frontopolar and ascending frontal regions respectively. The more anterior of these two areas of abnormal vascularization seemed to be supplied mostly by a branch of the middle meningeal artery.

Operation. On Nov. 3, 1954, a left frontal craniotomy revealed the presence of two large

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tumors embedded in normal cerebral parenchyma and closely adherent to the dura mater. The tumors were removed together with the dura mater, and the dural defect was repaired with temporal fascia.

Postoperative course was very good; however, at present, almost 3 years after her operation, the patient still shows some slight abnormalities in her behavior.

Microscopic Examination. The section consists of fibrocellular tissue, here and there dissociated by hemorrhagic areas. The cells are arranged in sinusoid streams running in different directions, richly anastomosed with one another. The nuclei are round or oval in shape, rich in evenly distributed chromatin and without nucleolus. The vascular component is well represented by capillary vessels or by larger blood spaces, the lining of which is often interrupted, showing extravasation of blood. In many areas the section consists of fibrocellular bundles arranged in whorls around a small vessel. The stroma appears formed by connective-tissue strands or by short fibrous tracts, partly showing hyaline degeneration (Fig. 1).

Case 2. No. 64/55. D.M., a 36-year-old male, brother of A.M., was admitted on Feb. 14, 1955. Five months previously he had a first generalized seizure with loss of consciousness during sexual intercourse; a similar episode occurred 1 month later in the same circumstances. Anticonvulsive therapy was prescribed, which he discontinued 1 month before admission, as seizures had not recurred. Ten days before admission he had a generalized seizure, another one followed in 24 hours, and he was admitted to our service.

Examination. He showed a slight left facial weakness and left hyperreflexia. Plain roentgenograms of the skull revealed no abnormalities. Mild bilateral peripapillary edema was present; visual acuity was 10/10 bilaterally. Cerebrospinal fluid pressure was normal, but protein content of the fluid was 80 mg. A right frontal focus was present on electroencephalogram. Percutaneous right carotid angiography showed a marked downward displacement of pericallosal and callosomarginal arteries.

Operation. On March 1, 1955, through a right frontal craniotomy, a large parasagittal
tumor, closely adherent to the dura mater, was exposed and removed. The dural defect was repaired with temporal fascia.

Postoperative course was uneventful and the patient is now back on his job as building supervisor. At present he has one seizure every 5 to 6 months.

Microscopic Examination. The section consists almost entirely of a layer of cells closely packed into cords running in different directions, and showing active proliferation. There is a loose connective stroma with a few blood vessels. The cells are polygonal with clear cytoplasm; the nuclei are large, unequal and vesicular, with chromatin in clumps. In a few marginal areas these elements merge into smaller ones, of more irregular shape, elongated, with smaller nuclei and denser chromatin. The loose connective stroma is partly hyalinized (Fig. 2).

Our cases, in regard to their microscopic structure, would seem to present certain similarities to those reported by Ectors and van Bogaert; however, while these authors were eventually able to find evidence of von Recklinghausen's disease in their patients, we have been unable, in spite of repeated and thorough examinations, to find such evidence.

The association of meningiomas and multiple neurofibromatosis is fairly frequent and the familial nature of von Recklinghausen's disease is well known. Therefore, the occurrence of meningiomas in members of families in which neurofibromatosis is present, even in its monosymptomatic forms, is quite feasible. The mechanism of this association is, nevertheless, a difficult one to explain, as the two tumors are derived from two different embryonic layers. Cushing and Eisenhardt, in their classic book on meningiomas relate the theory of Verocay, who thought that "... under the influence of some faulty development of the ectodermal portion of the nervous system, the adjoining mesodermal elements ... also have experienced a disturbance of development," and the hypothesis of Oberling, who thought that
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meningiomas might have to be considered lesions of neuro-ectodermal rather than mesenchymatous origin.

We have thought it worth while to report the cases of these 2 patients as they have so far yielded no evidence of harboring neurofibromas.

REFERENCES