Congenital Venous Malformation of the Scalp Associated with Plexiform Neurofibroma and Cranial Defect

Case Report

F. B. Maroun, M.D., F.R.C.S. (C), J. C. Jacob, M.D., F.R.C.P. (C), P. H. Markesteyn, M.D., and D. R. Mercer, M.D.

Department of Neurology and Neurosurgery, St. John's General Hospital, St. John's, Newfoundland, Canada

There are few reports of venous angiomas of the scalp, although references can be found to comparable lesions in the brain, spinal cord, skin, and skeletal muscle. The association of a venous malformation of the scalp with a plexiform neurofibroma and an underlying bony defect is rare and is the subject of this case report.

Case Report

A 21-year-old man had been aware of a painless swelling in the right posterior parietal area of the scalp since childhood. He had also noted a bluish discoloration of the skin and a defect in the skull underneath the swelling. There was no change in the size or consistency of this lesion until 2 months prior to admission when it suddenly grew larger and became tense. There was no history of preceding trauma. He promptly sought medical attention, and dark venous blood was aspirated from the lesion. He was then referred to this center for further evaluation. There was no family history of neurofibromatosis.

Examination. The blood pressure was 115/80 mm Hg and pulse rate 78 per min. Abnormal findings were confined to the head. The different components of the scalp lesion included venous lakes, peau d'orange appearance of skin, a tumor-like mass, and a hematoma (Fig. 1). Venous lakes were prominent when the patient was horizontal, but these collapsed quickly when the patient was placed in an upright position.

Routine blood analysis showed no abnormality. Chest x-ray film and electroencephalogram were normal. An electrocardiogram showed sinus rhythm, a cardiac rate of 60 per min, with an incomplete right bundle branch block pattern. X-ray studies of the skull (Fig. 2) showed an osteolytic lesion involving both the outer and inner table in the right parieto-occipital region and surrounded by a zone of dense cortical bone.

Percutaneous right carotid angiography showed normal arterial and capillary phases. In the venous phase the superficial and deep veins appeared normal, but the lateral sinus was markedly dilated. Attempts to catheterize the superficial temporal artery (for selective angiography) were unsuccessful, due to the marked narrowing of this vessel. The superficial temporal vein was large and easily catheterized. The resulting angiogram is illustrated in Fig. 3. This showed the presence of large venous channels draining into dilated venous lakes extending into the neck. The lateral sinus was not seen well, and the abrupt termination of one of the veins near

Received for publication January 21, 1969.
the bony defect suggested thrombosis and occlusion. External jugular vein catheterization and selective angiography showed filling of a large retromandibular vein but no communication with the large scalp veins.

Operation. The skin over the right lateral suboccipital and retromastoid areas was abnormal, being firm and dimpled over the swelling area (Fig. 4). The venous malformation was located in the galea, starting around the vertex, with large pools of venous blood draining into many channels and extending down through the bony defect to the lateral sinus. There was no arterial component to this lesion. In the suboccipital area a large “tumor” mass was present, measuring 6 × 6 cm; it was firm and adherent in part to the subcutaneous tissue. The bony defect measuring approximately 5 cm in diameter was located in the retromastoid area and seemed to have partly involved the mastoid air cells. Two large draining veins emptied into the lateral sinus. The dura was somewhat fibrotic along the bony defect. A large hematoma which was found adjacent to the “tumor” was evacuated. A total removal of the venous anomaly and the “tumor” was accomplished. The postoperative course was uneventful.

Histological Examination. The scalp and underlying soft tissue showed the presence of irregular masses of proliferating nerve sheaths (Fig. 5). In some areas these were arranged in bundles expanded by excess of both endoneural and perineural tissue show-

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Fig. 2. Osteolytic lesion involving inner and outer tables of the skull, in the right parietooccipital region. Note the zone of dense cortical bone surrounding the lesion.

Fig. 3. Selective angiogram after right superficial temporal vein catheterization. Large venous channels draining into venous lakes are seen extending down to the occipitomastoid region.

Fig. 4. Close-up of the skin lesion.