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

Case Report

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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.1,3 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

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Fig. 1. Components of the scalp lesion are outlined.

- a = the borders of the lesion
- s = peau d'orange appearance of skin
- h = hematoma
- v = venous lakes (arrows)
- e = infraauricular bluish discoloration of skin
- m = "tumor" mass (the plexiform neurofibroma)

The sutures indicate sites of superficial temporal and internal jugular catheterization.
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 showing mucoid degeneration and giving rise to a whorled and fascicular pattern. A more diffuse proliferation is seen in other areas. Numerous vessels, arteries, and veins as well as capillaries and large cavernous spaces were closely associated with the neurofibromatous tissue. In many areas there was hemorrhage around the proliferating nerve sheaths.

Discussion

The lesion described is a complex one consisting of a subcutaneous plexiform neurofibroma showing perineural hemorrhage, large subgaleal venous channels and lakes draining into an abnormal lateral sinus through a skull defect. Clinical and histological data suggested that this was a congenital lesion. The precise definition of the nature of
the venous lesion is important. Are these venous lakes and channels the result of compression by the plexiform neurofibroma on the lateral sinus with resulting stasis, dilatation, and eventual rupture? Or is the venous lesion a distinct congenital anomaly? For the following reasons we tend to favor the latter explanation:

1. The skull defect which was palpable, and the venous lakes which could be seen externally, had been noted since birth.
2. The presence of an endothelial lining would suggest a congenital lesion.
3. The lateral sinus was not occluded; on the contrary, it appeared markedly dilated.

In the course of normal development, the free communication between extracranial and intracranial venous circulation is obliterated except for small parietal and mastoid emissary veins, which pass through apertures in the cranial wall and establish communication between venous sinuses inside the skull and the veins external to it. Occasionally one of these connecting veins in the parietal or occipital areas remains unusually large. Under these circumstances, whenever intracranial pressure is decreased, there is a reversal of flow so that blood from intracranial venous sinuses enters the scalp and veins, and the latter become dilated and tortuous.

This type of anomaly should be differentiated from other vascular lesions involving the scalp, particularly the cirrhotic aneurysm. Detailed angiographic studies visualizing both external and internal carotid circulation, together with selective venous catheterization, should be performed to determine the nature, extent, and hemodynamic behavior of these lesions.

**Summary**

A complex congenital scalp lesion, comprising a subgaleal venous malformation, a plexiform neurofibroma, and an underlying skull defect, has been presented. Detailed angiographic studies have been shown emphasizing the purely "venous" nature of this lesion. The pathogenesis of the venous malformation and its relation to plexiform neurofibroma have been discussed.

**References**