4-year-old girl with a pink birthmark covering the right side of her face (Figure 1 and Figure 2) developed recurrent seizures. A plain roentgenogram of the skull (Figure 3) was obtained to support the suspected diagnosis.
Denouement and Discussion

Sturge-Weber Syndrome

Figure 1 and Figure 2. An extensive port-wine stain involves most of the right side of the child's face, extending onto the neck and shoulder.

Figure 3. Tear-track-like intracranial calcifications are present on a plain roentgenogram of the skull.

The most common vascular lesion associated with the Sturge-Weber syndrome is a port-wine stain or nevus flammeus, a vascular malformation present at birth and consisting of ectatic capillary to venu-ular-sized blood vessels in the dermis. The port-wine stain does not resolve over time, unlike capillary hemangiomas. There may be associated blood vessel abnormalities involving the eye on the same side of the face, which may subsequently lead to the development of glaucoma. Leptomeningeal angiomatosis on the ipsilateral surface of the cerebral hemisphere is the other major component of the Sturge-Weber syndrome, and this malformation is responsible for the development of seizures and mental retardation.

The distribution of the port-wine stain is of clinical importance in determining whether an affected child has an increased likelihood of having eye or central nervous system involvement. Areas of innervation of the trigeminal nerve are used to describe the distribution of the vascular patches on the skin of the face and are referred to as V₁, V₂, and V₃ areas for the branches of the fifth cranial nerve.

Seizures, reflecting leptomeningeal angiomatosis, were most likely to be present if the port-wine stain involved the V₁ area and other areas of the face, while patients developing glaucoma in association with a port-wine stain had at least the V₂ area involved.

Of the 274 patients in the study by Tallman et al. with port-wine stain involvement in the distribution of the trigeminal nerve, 22 (8%) had eye and central nervous system complications. All of these patients had port-wine stains involving the V₁ and/or V₂ areas of distribution. The upper eyelid in this study was considered to be part of the V₁ distribution and the lower eyelid part of the V₂ distribution. In addition, if patients had bilateral involvement, the incidence of eye or central nervous system complications was much higher. Although most patients with port-wine stains of the face will not develop seizures or glaucoma, involvement of the eyelids should alert one to the possibility of these complications, and ophthalmologic examination should be performed.

The soft tissue and bone underlying the port-wine stain may become hypertrophied, including the intracranial tissue involved with the stain. With increasing age, the port-wine stains have a tendency to become darker red or often purplish, and they may become nodular in areas.

PATHOLOGIC CORRELATION

The central nervous system vascular malformation present in Sturge-Weber syndrome is often confined to the pial vessels in the occipitoparietal area. The slow flow of blood through the malformed leptomeningeal vessels leads to progressive hypoxic injury, encephalomalacia, subsequent atrophy, and calcification in the cerebral cortex. These changes may result in seizures, intellectual impairment, and, less often, contralateral hemiparesis. Seizures are the most common neurologic disturbance, and they develop most commonly during the first year of life. The seizures are predominately focal motor or generalized and begin on the contralateral side of the body.

The most frequent ocular vascular anomaly associated with this syndrome is a hemangioma of the choroid. The conjunctivae frequently have dilated vessels. The development of glaucoma is thought to be the result of an abnormal plexus of episcleral vessels that results in increased intraocular pressure.

GENETICS

Sturge-Weber syndrome is sporadic in occurrence with no evidence of inheritance. There are no sex or racial predispositions. The exact incidence is unknown.

TREATMENT

Children with port-wine stains involving the eyelids should undergo careful ophthalmologic examinations and follow-up. Anticonvulsant therapy is required for the control of seizures. The flashlamp-pulsed tunable dye laser has proven to be the safest and most effective cosmetic treatment for port-wine stains. Children treated before age 7 years require fewer laser treatments to achieve the maximum lightening of the lesions.

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REFERENCES