

28. Tuberos Sclerosis and Sturge-Weber Syndrome

Tuberous sclerosis is an autosomal dominant (and spontaneously occurring) condition defined by facial adenoma sebaceum, seizures, and mental retardation. Intracranial findings are hamartomatous lesions consisting of subependymal nodules and cortical/subcortical tubers. Figure 28.1 A, B demonstrates multiple subependymal nodules lining the walls of the lateral ventricles. Although these lesions are clearly seen in this instance as low SI on T2WI (Fig. 28.1 A, white arrows) and high SI on T1WI (Fig. 28.2 B), the SI of these nodules varies (in part due to calcification), and in some instances the lesions are much less conspicuous. GRE better detects this calcification, which is absent in heterotopic gray matter—a lesion which potentially could be confused with the subependymal nodules of tuberous sclerosis, but that does not enhance and is isointense to gray matter on all pulse sequences. The enhancement of subependymal nodules is variable, although its presence on MRI (unlike on CT) is not indicative of malignant transformation to a subependymal giant cell astrocytoma (WHO grade 1). Distinguishing between these two lesions may thus be difficult and require close MRI followup. Fortunately, giant cell astrocytomas are slow-growing lesions with the major morbidity being from ventricular outflow obstruction. The FLAIR image in Figure 28.1 C demonstrates a subtle heterogenous area of high SI in the frontal horn of the left lateral ventricle that because of its location and size may represent a

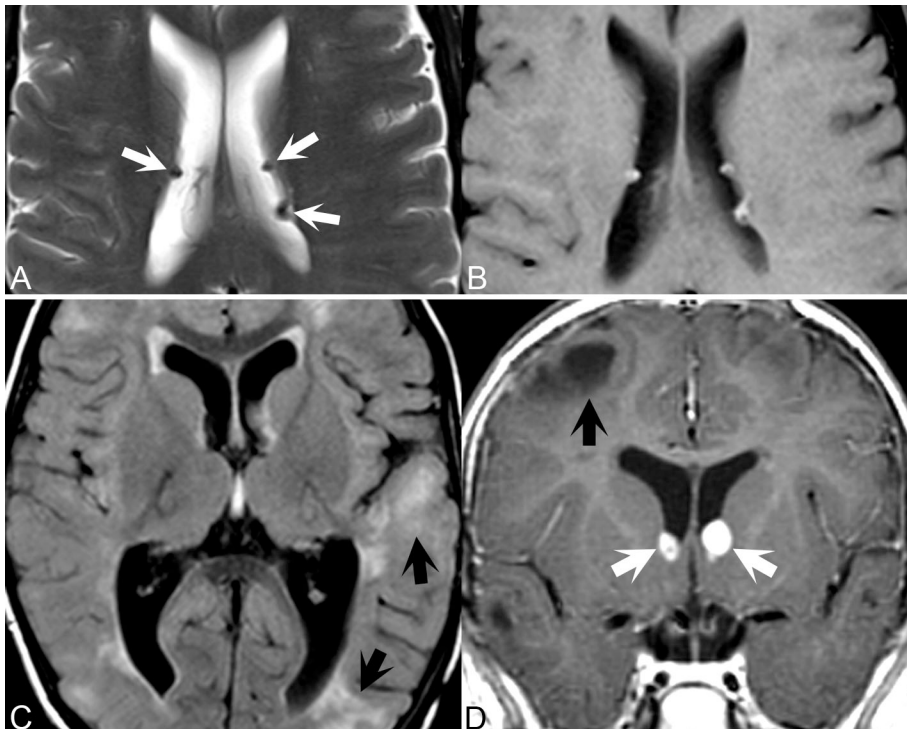


Fig. 28.1

giant cell astrocytoma. Coronal imaging in Figure 28.1 D reveals the existence of bilateral lesions (white arrows), that enhance and are both in the characteristic location—near the foramina of Monro—for a subependymal giant cell astrocytoma. Cortical and subcortical tubers occur most frequently in the frontal, then parietal, lobes and involve both gray and white matter. A “gyral core” pattern of tubers consists of an expanded gyrus surrounding a subcortical white matter hamartoma of high SI on FLAIR (Fig. 28.1 C, black arrows) and T2WI with low SI on T1WI (Fig. 28.1 D, black arrow). These lesions do not typically enhance. When two adjacent gyri are involved with sparing of the intervening cortex, a “sulcal island” pattern results. This is best seen on T2WI as two high SI subcortical lesions flanking a sulcal area of (normal) lower SI.

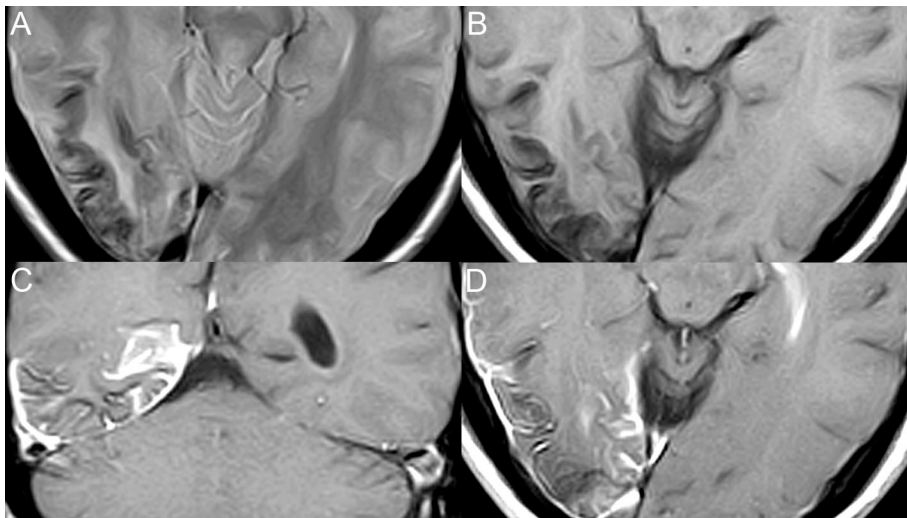


Fig. 28.2

Sturge-Weber syndrome is defined by the presence of a facial port-wine stain, mental retardation, and seizures. The port-wine stain is a capillary angioma, and ipsilateral to this lie the major intracranial pathologies of Sturge-Weber—cortical calcification, leptomeningeal angiomas, and parenchymal atrophy. Figure 28.2 A, B demonstrates a gyral pattern of abnormal low SI on T2 (A) and T1WI (B) that is most consistent with dense cortical calcifications. Contrast enhanced coronal (Fig. 28.2 C) and axial (Fig. 28.2 D) images display mild focal parenchymal atrophy along with leptomeningeal enhancement typical of the pial-contained thin-walled venous structures of leptomeningeal angiomas. Evolution of the vascular abnormalities can be seen, thought to be due to further occlusion of draining veins. In neonates, transient hyperperfusion may lead to pseudo-early myelin maturation which appears as decreased SI on T2WI. With subsequent ischemia and gliosis, increased SI on T2WI as well as enhancement may occur. An additional not uncommon finding is ipsilateral choroid plexus enlargement, together with prominent enhancement

Essentials of Clinical MR, 2nd edition

therein.