Sinus pericranii is a rare vascular anomaly involving abnormal communication between the intracranial and extracranial venous circulations. Although the condition can be diagnosed clinically, imaging is required to distinguish it from other entities and to identify the connection between the extracranial and intracranial dural venous sinus. We report the characteristic MR imaging and angiographic findings of a case of sinus pericranii.

Index words: Brain, arteriovenous malformations
Brain, sinuses
Brain, magnetic resonance (MR) angiography

Sinus pericranii is a vascular anomaly that usually occurs in the pediatric age group. Ohta et al. defined this condition as “a collection of nonmuscular venous blood vessels or venous hemangiomas adhering tightly to the outer surface of the skull bone and directly communicating with an intracranial venous sinus by way of many diploic veins of several sizes.” To our knowledge, few reports published in Korea have described the MR imaging and MR angiographic findings of sinus pericranii. We present a case in which the condition occurred in an infant, and in which the findings at imaging and histological examinations were characteristic.

Case Report

An 8-month-old boy, in whom a 2×2-cm soft, blue-tinged mass on the mid-line parietal scalp had been present since birth, was brought to our hospital. It was noted that coughing, crying, or assuming the head-down position caused the mass to increase in size. Physical examination showed that it was compressible, and neither tenderness nor pulsation was noted.

Doppler sonography of the scalp depicted a vascular mass with venous flow characteristics, while T1-weighted MR imaging demonstrated a mass of mixed hypo- and iso-signal intensity containing areas of signal void, indicating blood flow (Fig. 1A). The mass was heterogeneously hyperintense at T2-weighted imaging, and enhanced slightly after the intravenous administration of contrast material. Although MR angiography with a three-dimensional time-of-flight (3D-TOF) technique showed no arterial supply to the mass, 3D-TOF MR venography revealed a large emissary vein connecting the superior sagittal sinus with the dilated extracranial veins of the scalp (Fig. 1B). The anomalous vein was confirmed by digital subtraction angiography (Fig. 1C).

After ligation of the collateral veins of the scalp, en bloc resection of the mass and diploic veins was performed. Histologic examination confirmed that the mass comprised dilated anomalous veins lined with endothelial
Discussion

After the first related report by Hecker in 1845, Stromeyer described this anomaly as “a subperiosteal blood-filled cyst of the skull communicating with an intracranial sinus”, and proposed the term “sinus pericranii” (2). Although there are several synonyms for this anomaly, such as osteovascular fistula, venous tumor of the cranial bones, and subpericranial venous tumor, the term “sinus pericranii” has been generally accepted (3).

Three theories have been postulated regarding the possible pathogenesis of this anomaly: that it is congenital, spontaneous, and traumatic. Congenital theory as proposed by Muller contends that sinus pericranii is a true angioma coexisting with other congenital vascular anomalies such as venous angiomas, aneurysmal malformations of the internal cerebral vein, and cavernous hemangioma (3). The theory of spontaneous formation assumes either the development of a varix followed by pressure erosion of the skull, or chronic disease of the skull vault (3). Suggested pathogeneses of a traumatic sinus pericranii include fractures of the outer table of the skull, tearing of an emissary vein at the point of departure from the skull, direct sinus injury due to depressed fractures or bone fragments, and venous epidural hematoma due to sinus tearing (3-5). To determine its origin, histological examination is required; the nature of its endothelial lining permits differentiation between congenital and acquired types: while a congenital lesion is lined with endothelial cells, one that is acquired is lined with connective tissue (4). According to Arrues et al. (3), the spontaneous type of sinus pericranii is probably a variety of the congenital type.

Clinically, sinus pericranii most commonly occurs in patients under the age of 30, and tends to equally involve both sexes, particularly when congenital. Because of the higher incidence of head injuries among males, a slight male predominance has been reported in post-traumatic cases (6). The majority of cases of sinus pericranii occur at the midline, with its reported incidence varying according to location: 40% if frontal, 34% if parietal, 23% if occipital, and 4% if temporal (7).

The mass of sinus pericranii is soft and easily compressible, and increases in size when the patient’s head is lowered or when venous pressure is increased, such
as during the Valsalva maneuver or manual compression of the jugular vein. When pressure is direct, the mass disappears (5). Most patients are asymptomatic, but there are occasional complaints of local pain, vertigo, or headache (8).

A diagnosis of sinus pericranii is suggested by its clinical appearance, namely a fluctuant, bulging mass that varies in size and tension with changes in head position or intrathoracic pressure. Although definite diagnosis by means of the direct injection of contrast medium into the mass has been proposed, contrast-enhanced CT, MR imaging, or cerebral angiography usually yields enough information to make a diagnostic decision (7, 8), and conventional radiographs can demonstrate a well-defined bony defect in the area of the mass. Color Doppler ultrasound depicts a blood pool within the mass, with venous flow revealed by pulsed Doppler study (9). Contrast-enhanced CT visualizes the venous filling within a scalp mass, as well as an associated skull defect, and cerebral angiography reveals a vascular scalp lesion which communicates with the superior sagittal sinus through the dilated emissary veins. Sometimes, however, it is difficult to demonstrate at angiography the existence of venous communication, especially in cases in which venous flow is very slow and weak (3). By virtue of its superior contrast resolution, high sensitivity and multiplanar capability, MR imaging demonstrates the characteristic appearance of sinus pericranii, namely a soft-tissue mass of mixed signal intensity containing areas of signal void, as in the present case. MR venography, furthermore can provide vivid evidence of communication between the extracranial and intracranial dural venous systems, and is likely to be used as an alternative to conventional angiography (3). In the present case, MR venography appeared to be comparable to conventional angiography. Although the emissary vein was slightly clearer at conventional angiography, excellent depiction of the course and relations of the intracranial and extracranial veins was demonstrated equally by both techniques.

The differential diagnoses of sinus pericranii include congenital or traumatic arteriovenous communication, post-traumatic subepicranial varix, venous cavernoma, meningo (encephalo)cele, cephalohematoccele, eosinophilic granuloma, and epidermoid tumor (4, 5).

Although spontaneous regression has been reported, most patients with sinus pericranii require removal of the sinus and the ligation of communicating veins. Indications for surgery include cosmetic problems and the prevention of massive hemorrhage and traumatic air emboli (3, 5, 10).

References

2008:49:417-420