The Neuro Image Quiz, Answers

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This child has a choroid plexus cyst.

Cysts of the choroid plexus are very common and are rarely symptomatic. Usually they are incidental findings on sonograms of the fetal or neonatal head [1]. In several post mortem studies, choroid cysts have been found in more than 50% of routine specimens [2, 3]. The majority of these choroid cysts were less than 1 cm in size. They occur equally in all age groups with no sex predilection.

Choroid plexus cysts usually remain small and static in size or regress spontaneously. Chudleigh et al. [4] reported 5 cases of transient choroid plexus cysts that resolved spontaneously in utero. Ultrasound documented the presence of these cysts in fetuses of less than 20 weeks gestation. Follow-up sonograms documented spontaneous disappearance of the cysts between 20 and 23 weeks gestational age. In each case the baby was neu-rologically normal at term; neonatal neurosonograms showed no pathology.

Rarely, choroid plexus cysts may grow sufficiently large to cause symptoms. To our knowledge, only 8 such cases have been reported in the English language literature. The 8 patients ranged in age from 7 months to 64 years. Five of the 8 were 10 years old or less. These unusually large cysts ranged from 1.5 to 9 cm in diameter and typically were centered on the trigone of the lateral ventricle. Such cysts may evaginate from the lateral into the third ventricle. Giant choroid cysts arising primarily within the third and fourth ventricle have not yet been reported.

Clinically, the vast majority of patients with small choroid plexus cysts remain asymptomatic and require no therapy. The few patients with giant choroid plexus cysts present with signs of intermittent increased intracranial pressure, i.e. headache, focal or generalized seizures, papilledema and visual disturbances. The elevation of intracranial pressure is believed to be caused by either (a) transient obstruction of the foramen of Monro and/or (b) trapping of the temporal horn of the lateral ventricle.

Radiologically, choroid plexus cysts appear as thin-walled fluid collections. Small choroid plexus cysts appear on ultrasound as fluid-filled hypoechoic lesions within or projecting from the choroid plexus. The cyst wall may appear thick or thin, depending on how deeply the cyst is buried in the choroid. The small cysts are

usually observed near to the glomus or near to the foramen of Monro.

The giant cysts appear as focal dilatations of one lateral ventricle. These may cause, or mimic, unilateral hydrocephalus. On CT, the cyst wall is usually very smooth and thin with no calcification and no appreciable enhancement of the cyst wall. The choroid plexus from which the cyst arises is usually displaced by the cyst and is draped over one arc of the cyst wall after contrast administration.

Histologically, the cyst wall is composed of a thin, loose network of connective tissue that is lined by a single layer of flattened, unciliated epithelial cells. No glial elements are present. The cyst is firmly attached to an otherwise normal choroid plexus at a single point and is easily separable from the ependymal lining of the ventricle. The fluid within the lumen is clear, acellular and chemically indistinguishable from CSF.

Choroid plexus cysts are believed to be developmental lesions. The fetal choroidal villi are formed by infolding of the choroidal epithelium into the choroid matrix [2]. Some of the epithelial cells at the apex of the infolding may detach from the remainder of the epithelium to form tubuli or small cysts. Subsequent accumulation of epithelial secretions within the closed space is believed to increase the size of the cyst.

Inflammation is well known to cause cyst formation in other tissues. Subependymal cysts commonly result from viral infection such as rubella and cytomegalovirus [5]. At least some of the choroid cysts may be expected to be of inflammatory origin.

Radiological differential diagnosis of the thin-walled intraventricular cysts includes cysts of other histology such as ependymal cysts, arachnoid cysts and intraventricular cysticercous cysts. Pseudoloculation of the ventricles by juxtaventricular inflammatory cysts might also have similar appearance [6].

Practically, it is difficult to distinguish the giant choroid plexus cyst from other intraventricular cysts. The cysticercous cyst is probably the most common large, symptomatic intraventricular cyst. Serologic testing will determine whether the patient has cysticercosis and will suggest that any cyst detected is, or is not, a cysticercous cyst. Paraventricular tumors such as thalamic gliomas may develop large cysts that bulge into the atri-

um. Such cysts may even lie adjacent to the choroid plexus. However, these tumor cysts are usually distinguishable by the associated parenchymal mass. Those familiar with choroid plexus cysts will usually have little difficulty in recognizing them radiologically and in understanding their benign nature.

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