The Neuro Image Quiz, Answers

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We will define a line of union of pia with ependyma as a ‘pial-ependymal seam’. We will define a ‘lip’ of gray matter as a layer of gray matter that lies immediately adjacent to a pial-ependymal seam and that forms a continuous column of gray matter between the deep (ventricular) surface of the brain and the superficial (pial) surface of the brain. Schizencephaly may then be defined as a congenital disorder of neuronal migration that is characterized by the presence of unilateral or bilateral columns of heterotopic gray matter containing a pial-ependymal seam. If the columns of gray are juxtaposed to each other immediately to each side of the pial-ependymal seam, the condition is designated schizencephaly with closed lips. If the columns of gray are widely separated by an intervening diverticulum of ventricular CSF, then the condition is designated schizencephaly with open lips. In this 9-month-old girl, the columns of gray (white arrowheads in fig. 1 in The Neuro Image Quiz, Questions) are widely separated, indicating schizencephaly with open lips. Answer 1 is false. Answer 2 is false. Schizencephaly is believed to develop in the second month of gestation when a focal insult to the primitive brain wall prevents a smaller or larger portion of the wall from differentiating into the three (matrix, mantle and marginal) layers of His. In this dysgenetic zone, brain growth is retarded. The cerebral wall remains thinner and is dimpled on both its superficial and deep sides. On the deep, ventricular surface of the zone, the single-layered ependyma thickens to multiple layers and extends outward into the dimple. On the superficial, pial surface of the zone the pia matter extends inward into the dimple. In the depths of the dimple the pia becomes continuous with the ependyma, completing the pial-ependymal seam. Because this zone does not differentiate into the three layers of His, neu-roblasts mature into gray matter throughout the zone. This creates a continuous column of gray matter that extends through the full thickness of the brain wall around the pial-ependymal seam.

If the zone of insult is relatively small and if there is no hydrocephalus, then the pial-ependymal seam is believed to remain narrow. The columns of gray appose each other closely, so the condition is designated schizencephaly with closed lips. Should the zone of insult be larger and should there be concurrent hydrocephalus, then expansion of the ventricle creates a diverticulum of CSF that bulges the dimple outward, stretches it and separates widely the ap-posing lips of gray matter. This is schizencephaly with open lips. All around the edges of the defect, the pia still merges with the ependyma and forms a raised fringe designated the pial-ependymal ridge (fig. 1). The original pial-ependymal seam becomes stretched out over the diverticulum like a drumhead that attaches circum-ferentially to the pial-ependymal ridge. This highly streched, pial-ependymal seam is designated the pial-ependymal membrane or the ‘roofing’ membrane, since it roofs over the diverticulum. With marked hydrocephalus, the diverticulum may bulge remarkably and even evaginate between the cortex and dura. The overlying hemicalvarium is often thinned. These findings should be understood as components of open lip schizencephaly and should not be mistaken for arachnoid cyst or other extraaxial collections. Answer 3 is false.

Two criteria must be met for accurate diagnosis of open lip schizencephaly: (a) the presence of continuous full-thickness transcerebal columns of heterotopic gray (the lips) (white and black arrowheads in fig. 1 in The Neuro Image Quiz, Questions and fig. 1. and (b) separation of those lips by a full-thickness, transcerebral ventricular diverticulum filled with CSF [1–6]. Answer 4 is D (A and B). The septum pellucidum is commonly absent (67%), as is the corpus callosum (20%), but these are accessory features not intrinsic features of schizencephaly. The open-lip form of schizencephaly is more common (72%) than schizencephaly with closed lips (28%). Open-lip schizencephaly is usually bilateral (78%). Closed-lip schizencephaly is bilateral in 43% and unilateral in 57%. There is no sex predilection. Most schizencephaly is discovered in childhood (mean age – 6 years), but occasional cases escape detection until late, e.g. age 53 years.

In patients with schizencephaly the head is commonly asymmetrical. With unilateral schizencephaly, the head is asymmetrical in 88% of cases. In unilateral closed lip schizencephaly the side of the schizencephaly

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Fig. 1. Pathological specimen of schizencephaly with open lips. matter (black arrowheads) that line the diverticula. At the superficial edges of the gray columns, the pia and the ependyma merge into a pial-ependymal ridge (crossed white arrow) that gives rise to the pial-ependymal roofing membrane (white arrows). On the right, the open lips can be seen to close together posteriorly. c Coronal section through the posterior part of the third ventricle shows narrowing of the lips on the left and fusion of the lips into a closed-lip schizencephaly (black arrowheads) on the right. Islands of heterotopic gray matter are present bilaterally (black arrows). tinuous, full-thickness transcerebral columns of heterotopic gray

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is asymmetrically smaller in each case (100%). In unilateral open lip schizencephaly the ipsilateral side can be larger (50%), smaller (25%) or equal (25%) in size to the contralateral side.

In bilateral schizencephaly the head is asymmetrical less commonly (59%). In bilateral closed lip schizencephaly, the side of the larger defect can be larger (67%) or equal (33%) to the contralateral side, not smaller. In bilateral open lip schizencephaly, the side of the larger defect can be larger (73%), equal (20%) or smaller (7%) than the opposite side. Indeed, the size of the hemispheric ventricle appears to vary with the size of the ventricle and can be substantially altered by shunting the diverticulum.

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