Latin Comparative Pathology Group
The Latin Subdivision of the CL Davis Foundation
Diagnostic Exercise

Case #: 50  Month: November  Year: 2014

Answer Sheet

Contributor: M. Kelly Keating, DVM, DACVP, Case courtesy of the Comparative Pathology Laboratory, University of California–Davis, CA.

Clinical History: A 1-month-old Lhx6<sup>−/−</sup> mouse on Balb/c strain background presented with apparently non-painful swellings on the head, but otherwise bright, alert, and within normal limits.

Diagnosis: Bilateral encephaloceles and cranioschisis with ventriculomegaly and multiple intra-parenchymal cavities.

Description from gross examination: Two bilaterally symmetric, soft to fluctuant, 4–5 mm in diameter swellings elevate the skin overlying the frontal bone (image 1). On coronal section, the swellings correspond to defects within the calvarium, and subcutaneous cavities that are contiguous with the cerebral vault (images 2 and 3).

Image 1.
Differential diagnoses based on image 1 & microscopic findings:

- **Encephalocele/ Meningoencephalocele:** Herniation of neural tissue and meninges through a defect in the skull

- **Meningocele:** Herniation of meninges through a defect in the skull

- **Lipomeningocele:** Similar to a meningocele, but with large amounts of associated adipose tissue

- **Subcutaneous mass:**
  - **Hematoma:** Collection of hemorrhage and often fibrin, fibrosis, mixed inflammatory cells, and hemosiderin/hematoidin
  - **Lipomatous hamartoma:** Solitary mass on midline composed of adipose and fibrous stroma that expands the subcutis and can extend into the calvarium and ventricular system

**Microscopic findings:** Two bilaterally symmetric exophytic cysts protrude through defects in the frontal bone, elevate the overlying dermis and haired skin, and are lined by ciliated epithelium (ependyma) and an external rim of fibrous connective tissue (images 3-5). The cysts are contiguous with markedly dilated and distorted lateral ventricles, and there is variable attenuation of the cerebral cortex. Multifocally there are small cavities within the cerebral cortex, one of which communicates with the lateral ventricle. The cavities are not lined by ependyma, are delineated by spongiotic neural parenchyma and contain clear space and small numbers of erythrocytes. In the subcutis and fascia surrounding the protruded neural tissue are small numbers of lymphocytes, plasma cells and macrophages.
Image 4. HE. Subgross

Image 5. HE.
Discussion:

The swellings are consistent with two bilaterally symmetric encephaloceles, associated with marked dilation and distortion of the lateral ventricles and several intra-parenchymal, periventricular cavities. In this case these changes are consistent with congenital anomalies based on age of onset and lack of evidence for an infectious or traumatic etiology. Multiple congenital malformations in association with the presence of an encephalocele have been commonly reported. Both encephaloceles and primary ventriculomegaly are typically associated with abnormal neural tube development. In laboratory mice, encephaloceles have been rarely reported in the literature, and when reported are often in association with in utero death or experimental manipulation (radiation).

References and Recommended literature:


Please send your comments/questions to the whole LCPG list by hitting “reply to all”.

A final document containing this material with answers and a brief discussion will be posted on the C. L. Davis website by the end of the current month (http://www.cldavis.org/lcpg_english.html).