

SECTION EDITOR: ENID GILBERT-BARNES, MD

## Pathological Case of the Month

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**A** 1-YEAR-OLD BOY had a history of fever, poor feeding, lethargy, and irritability for 1 month. Computed tomographic scans of the head revealed a right infratentorial hyperdense irregular cystic mass with hydrocephalus (**Figure 1**).

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Cerebrospinal fluid revealed no leukocytes; protein, 40 g/L; glucose, 4.4 mmol/L (blood glucose, 5.5 mmol/L); and negative findings from Gram stain and sterile culture. A clinical diagnosis of tuberculous meningitis with tuberculoma and hydrocephalus was made. Shunt surgery was performed, but the infant had a cardiac arrest. A brain autopsy was performed (**Figures 2, 3, and 4**).

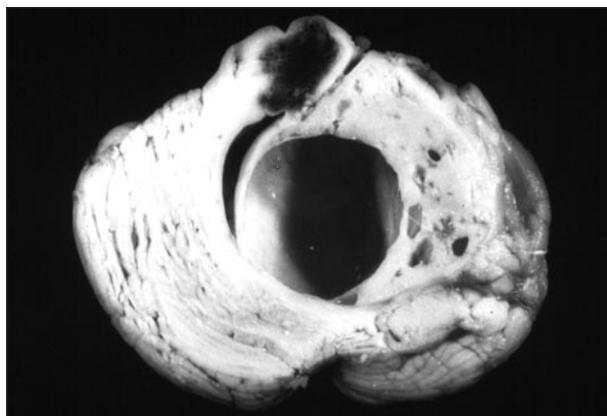


Figure 1.

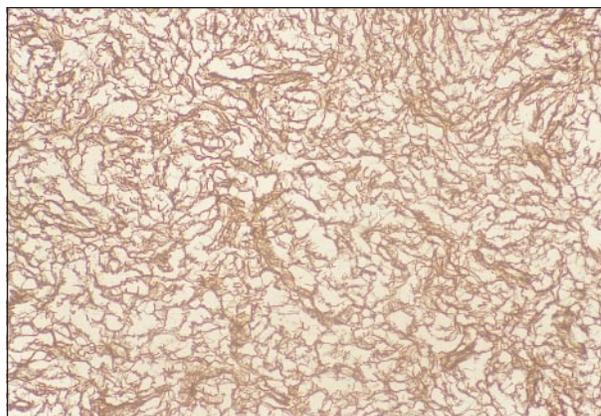


Figure 3.

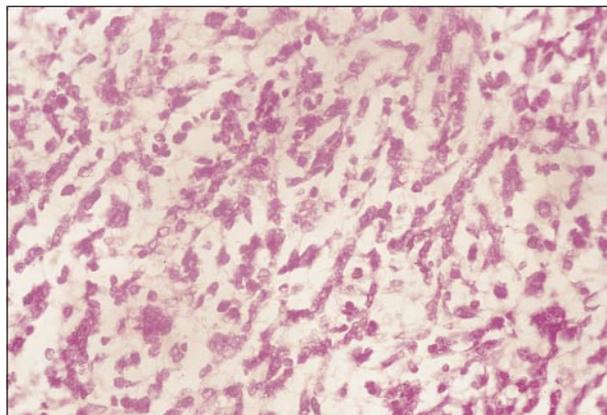


Figure 2.

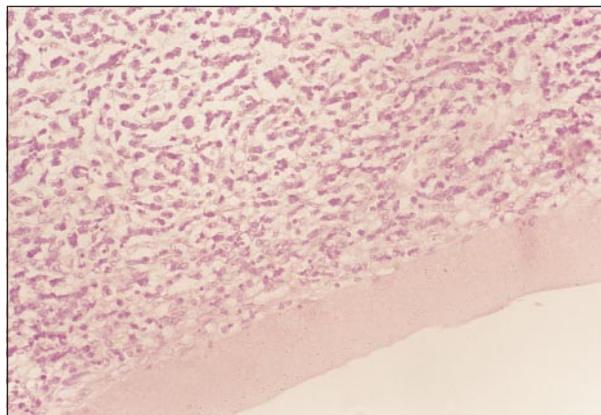


Figure 4.

# Diagnosis and Discussion

## Cystic Desmoplastic Medulloblastoma of Infancy

**Figure 1.** Cystic tumor (6 × 5 × 4.5 cm) in the right cerebellum. The largest cyst measures 3 cm in diameter and is filled with greenish fluid.

**Figure 2.** Undifferentiated areas composed of cells with small hyperchromatic nuclei and inconspicuous cytoplasm. The cells are arranged in cords and trabeculae (hematoxylin-eosin, original magnification ×280).

**Figure 3.** Marked desmoplasia with connective tissue encircling single cells or groups of cells (reticulin stain, original magnification ×550).

**Figure 4.** Scattered ganglion cells in a fibrotic background (hematoxylin-eosin, original magnification ×280).

**A**t autopsy, the brain weighed 955 g. The right cerebellum contained a cystic tumor measuring 6 × 5 cm. Medulloblastomas are dense small cell tumors. They are heterogeneous but as a group are known for small hyperchromatic condensed nuclei and inconspicuous cytoplasm. There are several classifications: undifferentiated, neuroblastic differentiation, glial differentiation, mixed glial neuronal differentiation, and divergent differentiation.<sup>1</sup> The desmoplastic variant of cerebellar medulloblastoma<sup>2</sup> formerly identified by some workers as an arachnoidal cerebellar sarcoma<sup>3-5</sup> is a superficial well-circumscribed form of medulloblastomas that predominantly occurs in young adults. Desmoplastic medulloblastoma has less nodularity compared with the other counterparts and a higher content of reticulin surrounding single cells or groups of cells. Abundance of connective tissue lends firmness to the tumor. The unique features of this case include (1) multiple cysts; (2) marked desmoplasia in a 1-year-old child; and (3) three types of clear-cut areas with abrupt transition: undifferentiated areas, mesenchymal areas with ganglion cells, and transition areas with follicular architecture (ie, pale islands).

Differential diagnosis of this tumor includes other cystic tumors and morphologically, desmoplastic tumors. Other differential diagnoses include (1) desmoplastic cerebral astrocytoma of infancy, (2) desmoplastic infantile ganglioglioma, (3) pilocytic astrocytoma of cerebellum, and (4) hemangioblastoma.

Desmoplastic cerebral astrocytoma of infancy<sup>6-8</sup> is a superficially situated, supratentorial glioma. Grossly, the lesion appears as a large, firm, globular mass, densely adherent to the surface of the brain. A distinctive feature is the intense desmoplasia that imparts a fibrotic appearance. Scattered elongated glial cells, strongly positive for glial fibrillary acid protein, are seen. No ganglion cells are seen. Desmoplastic infantile ganglioglioma<sup>9,10</sup> are supratentorial, superficially situated cystic tumors. Microscopically they show collagenous rich tissue of varying cellu-

larity with an appearance of a neurofibroma. Ganglion cells are plump, small to medium sized, and mixed with glial cells or isolated within the desmoplastic tissue. The neuronal nature of these cells is suggested by their roundness, vesicular nuclei, variably prominent nucleoli, and Nissl substance. The spindle cells are strongly positive for glial fibrillary acid protein.

Pilocytic astrocytoma of the cerebellum can be seen as a mural nodule within a cyst. Despite gross circumscription, they permeate the surrounding tissue, particularly the white matter for some distance involving the overlying leptomeninges. They occur in the second decade of life. Microscopically stellate astrocytes in microcystic regions, Rosenthal fibers, and eosinophilic granular bodies are seen.

Hemangioblastomas, another cystic tumor of the cerebellum, are seen in adults and are dark red or yellow. A network of dilated vessels with variably lipidized stromal cells is seen microscopically. This tumor morphologically showed 3 types of clear-cut areas: undifferentiated areas, fibrotic areas with ganglion cells, and transition areas with follicular architecture (ie, pale islands) along with multiple cysts and marked desmoplasia.

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### REFERENCES

1. Burger PC, Grehmann PC, Bliestle A, Kleihues P. Differentiation in the medulloblastoma: a histological and immunohistochemical study. *Acta Neuropathol.* 1987; 73:115-123.
2. Rubinstein LJ, Northfield DWC. The medulloblastoma and the so-called "arachnoidal cerebellar sarcoma": a critical re-examination of a nosological problem. *Brain.* 1964;87:379-412.
3. Foester O, Gagel O. Das umschriebene arachnoidal Sarcom des Kleinhirns. *Z Gesamte Neurol Psychiatr.* 1939;164:565-580.
4. Langheim W, Kernohan JW, Uihlein A. Arachnoidal sarcoma of the cerebellum. *Cancer.* 1962;15:705-716.
5. Ramsey HJ, Kernohan JW. Circumscribed sarcoma of the cerebellum: an electron microscopic study. *J Neuropathol Exp Neurol.* 1964;23:706-718.
6. Taratulo AL, Monges J, Lyeyk P, Leiguarda R. Superficial cerebral astrocytoma attached to dura: report of six cases in infants. *Cancer.* 1984;54:2502-2512.
7. de Chadarevian JP, Pattisapu JV, Faerber EN. Desmoplastic cerebral astrocytoma of infancy: light microscopy, immunocytochemistry and ultrastructure. *Cancer.* 1990;66:173-179.
8. Aydin F, Ghatak NR, Salvant J, Muizelaar P. Desmoplastic cerebral astrocytoma of infancy: a case report with immunohistochemical, ultrastructural and proliferation studies. *Acta Neuropathol (Berl).* 1993;86:666-670.
9. Ng THK, Fung CF, Ma LT. The pathological spectrum of desmoplastic infantile gangliogliomas. *Histopathology.* 1990;16:235-241.
10. Vanderberg SR, May EE, Rubinstein LJ, et al. Desmoplastic supratentorial neuroepithelial tumors of infancy with divergent differentiation potential ("desmoplastic infantile gangliomas"). *J Neurosurg.* 1987;66:58-71.