Pathologic Analysis of CNS Surgical Specimens

Peter C. Burger, MD

Interdisciplinary Quality Control

- Familiarity with entities
- Use of diagnostic algorithm
- Consideration of clinical, radiologic and pathologic features
- Communication
- Consultation
- Education
- Follow-up

Intraspinal Lesion In An Adult

Immunohistochemistry for EMA
Calcifying pseudoneoplasm of the neuraxis

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Diagnostic Algorithm
- Is there a lesion?
- If so, is it neoplastic
- If neoplastic, what tumor type?
- What grade?
- Is pathological diagnosis c/w clinical and radiologic findings?

Case
- 18 year-old woman
- Headaches, vomiting, blurred vision
- Obstructive hydrocephalus
- Cystic pineal region mass

Calcified, Cystic Pineal Region Mass

Diagnosis?
Pineal Region Neoplasms

- Pineal parenchymal tumors (pineocytoma, of intermediate differentiation, pineoblastoma)
- Papillary tumor of the pineal region
- Other regional, e.g. pilocytic astrocytoma, ependymoma

Normal Tissues Potentially Misinterpreted as Abnormal

In Frozen Sections

- Compressed normal hypomycotinal vessels as vascular malformation
- Gray matter as necrosis
- Normal perineuronal oligodendroglia as infiltrating tumor cells
- Corpus striatum and thalamus with "pencil fibers" of white matter as malformation or neoplasm
- White matter as gliomas
- Collateral bulb as neoplasm
- Fasiculi dentati of the hippocampus as a neoplasm or malformation
- Cerebral amylic as infectious organisms
- Contaminating "bone dust" as tissue calcification
- Choroid plexus as choroid plexus papilloma

- Pineal gland as pineal parenchymal tumor or other neoplasm
- Crushed cells in adenohypophysis as small cell neoplasm
- Neurohypophysis as neoplasm (pituitary, pilocytic astrocytoma)
- Intermediate lobe cyst as Rathke cleft cyst
- Cerebellar internal granular cell layer as small cell neoplasm
- Cerebellar external granular cell layer in an infant for subependymal tumor aggregation
- Ependymal cells of spinal cord as intramedullary neoplasm
- Sensory ganglia (e.g. dental root) as intraneural neoplasm

Fig. 3.26. Papillary Tumor of the Pineal Region. This neoplastic focus should be considered in the face of a diagnosis, enhancing pineal region mass. Pineal parenchymal tumors, germinomas, and pituitary are some common entities in this region.
Normal Pineal With Brain Sand

Normal Pineal

Synaptophysin

PPTID vs. Pineoblastoma

Papillary Tumour of the Pineal Region

Normal Pineal !

Calcified, Cystic Pineal Region Mass – Pineal Cyst
Pineal Cyst

- Young adults, more often women
- May be obstructive, with hydrocephalus, HA, vomiting, etc.
- Often asymptomatic, incidental finding

Normal pineal gland in pineal cyst

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College Student With T2-Bright Frontal Lobe Lesion:

Clinical Diagnosis - “B.T.”

“Mixed Glioma (Oligoastrocytoma)”
Demyelinating Disease (Multiple Sclerosis)

Astrocytoma Grade II

Demyelinating Disease

Oligodendrogioma-Like Lesions
- Oligodendroglioma
- Diffuse astrocytoma
- **Macrophage-rich lesions**
- Clear cell ependymoma
- Clear cell meningioma
- “Neurocytic” tumors
  - Neurocytomas
  - Dysembryoplastic neuroepithelial tumor
  - Rosette-forming glioneuronal tumor (of the fourth ventricle)
Demyelinating Disease - Histological Features

- White matter localization
- Sharp border
- Macrophage infiltrate
- Lymphocytic infiltrates (some cases)
- Preserved axons
- Reactive astrocytes
- Creutzfeldt cells
Hypercellularity, Sharp Border

Demyelinating Disease – Macrophages (CD68)

Demyelinating Disease – Neurofilament Protein (Preserved Axons)

Demyelinating Disease – Macrophages and Creutzfeldt Cell

Demyelinating Disease – Macrophages and Creutzfeldt Cells

Demyelinating Disease – Creutzfeldt Cell and Granular Mitosis
Consequences of Misdiagnosis of Demyelinating Disease

- Psychological trauma ("pain and suffering")
- Biopsy, esp. when open, resulting in neurological deficit
- Attempt at excision after frozen section diagnosis of tumor, resulting in neurological deficit
- Radiotherapy with radionecrosis

Demyelinating Disease - Radiologic Features

- Solitary or multiple
- Any location, but lesions approached surgically usually subependymal, subcortical, or spinal intramedullary
- Considerable edema in some cases
- "Open ring" or "horseshoe pattern" of CE

"Trust, but verify!"*

* Ronald Reagan

Normal Cerebellum
Diagnostic Algorithm

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Pilocytic Astrocytoma with Vascular Proliferation

Radiologic Features Suggesting Low Grade Lesion

• Skull erosion
• Lack of mass effect in contrast-enhancing lesion
• Absence of perilesional edema in contrast-enhancing lesion
• Dense calcification
• Macrocyst
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Demyelinating Disease - “Open Ring” Configuration

“Open ring” pattern of contrast enhancement

Inconsistency:

Grade II infiltrating astrocytomas and oligodendrogliomas rarely enhance

Image inconsistent with grade II infiltrating glioma
Multiple Sclerosis

Cerebral Deep Venous System (Postmortem “Venogram”)

Sites and Patterns of Contrast Enhancement

- **Dura**
  - Focal
  - Neoplasms, e.g. meningioma
  - Non-neoplastic lesions, e.g. sarcoidosis; Rosai-Dorfman disease
  - Diffuse and bilateral – result of intracranial hypotension, with layer of proliferating vessels on inner aspect of the dura
- **Leptomeninges**
  - Focal
  - Diffuse (inflammatory lesion; disseminated primary or secondary neoplasm)
- **Parenchyma**
  - “Ring” or “rim” pattern – glioblastoma, primary CNS lymphoma in immunocompromised patients; abscess; some metastases; some infarcts
  - “Open ring” – demyelinating disease
  - Solid (“homogeneous”) – most compact; discrete tumors, e.g. pilocytic astrocytoma; ependymoma, sporadic primary CNS lymphoma
  - Cortical, gyriform – infarct; inflammatory disease, e.g. herpes simplex encephalitis, radionecrosis

Enhancing Edema-Producing Lesion

Is this specimen representative?

NO!
Is this specimen representative?

Non-enhancing Region of “Edema”

With contrast

FLAIR

Is this specimen representative?

Maybe.

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Radiologist-Clinician-Pathologist Communication

- Unscheduled/informal
- Tumor board or equivalent
- Pathologist in OR

Pathologists’ View of Neurosurgeons
If, as a pathologist, you are asked, or even if only slightly encouraged, to get an outside consultation on one of your cases, the correct response is:

A. Ignore it
B. Tell him/her you are confident of your impression and a consultation is not necessary
C. Sulk, passive-aggress and send it with great reluctance and delay
D. “Of course, I’ll send it out today”

Examples

Brain Tumor In A 65-year-Old
Brain Lesions in A 65-Year-Old - Pre- and Post Steroids

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Older patient

Response to steroids

Solid enhancement

Multiplicity

PCNSL

Primary CNS Lymphoma Treated With Steroids
52-year-old Man with Intramedullary Spinal Cord Tumor

Discrete, Enhancing Intramedullary Tumor

Well circumscribed*
- Pilocytic astrocytoma
- PXA
- Ependymoma
- Choroid plexus tumors
- Astroblastoma
- Ganglion cell tumors
- Hemangioblastoma
- Meningioma

Infiltrative
- Diffuse astrocytomas (grades II, III, IV)
- Oligodendrogliomas (grades II and III)
- Primary CNS lymphoma
- ?? Medulloblastoma

* Extent of resection the most important prognostic factor; most contrast enhance

Intramedullary Tumors by Degree of Circumscription

Discrete, Highly Vascular Intramedullary Tumor

Well Circumscribed Intramedullary Tumors
- Ependymoma
- Hemangioblastoma
- Pilocytic astrocytoma
- Schwannoma (rare)
- Paraganglioma (filum)

Intramedullary Tumor - Frozen Section
Diffusely Infiltrative Astrocytoma

Spinal Intramedullary Tumors - Surgical Options

- Diffuse astrocytoma - stop (biopsy only)
- Discrete lesion - resect, with risk of producing neurological deficit

Intramedullary Tumor With Discrete Border – “Frozen”

Intramedullary Tumor

Frozen section

Oil-red-O

Hemangioblastoma

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