The Many Faces Of Intracranial Lymphoma

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Learning objectives

1) To discuss epidemiology and imaging of both primary and secondary lymphomatous involvement of the brain and revisit useful imaging signs.

2) To outline the differences and overlaps in lymphoma imaging in both immunocompromised and immunocompetent patients.

3) To discuss potential pitfalls in neuroimaging of intracranial lymphoma.

4) To discuss role of advanced imaging modalities as problem solving tools.
Background

Primary Central Nervous System Lymphoma (PCNSL), by definition, is limited to the brain, leptomeninges, spinal cord or eyes without any evidence of spread outside the CNS at time of diagnosis [1].

PCNSL occurs in about 6% of patients with AIDS and is now believed to be more common than low grade Astrocytomas and as common as meningiomas [2]. Congenitally immunodeficient patients and those who are immunosuppressed post organ transplantation are also predisposed [2, 3, 4].

The incidence of PCNSL in immunocompetent patients has also increased over the past two decades [1, 2]. Currently, PCNSL accounts for 1-5 % of all brain tumors and about 1 % of all NHL's [1, 5, 6].

Secondary lymphomatous CNS involvement can also occur and may be indistinguishable from PCNSL on imaging.

Pathologically, PCNSL is invariably of the Non Hodgkin (NH) subtype. Most of the cases are composed of large lymphomatous cells with a B cell phenotype [1, 4].
Imaging findings OR Procedure details

From a radiological perspective, CNS lymphomas are great imitators. Although some imaging findings may be suggestive, none are pathognomonic [6].

PCNSL in Immunocompetent Patients:

It usually presents in the sixth decade. Lesions are mostly solitary (up to 80%), with multifocal lesions reported in 20-40% of patients [1, 5].

Most lesions involve central hemispheric or periventricular white matter (Fig-1). Proximity to the sub arachnoid/subependymal space is a common finding and may provide a diagnostic clue [7].

Frontal lobe involvement is seen in 20-43% of cases. Other sites of involvement include basal ganglia (13-20%), corpus callosum (10%), posterior fossa (9-13%) and spine (1-2%). Most lesions are iso dense (46%) to hyper dense (46%) on non contrast CT scans [5].

On T2W images, the lesions are often hypo intense to grey matter [1]. Lesions are usually hyper intense on DW images (Fig-2) and hypo intense on ADC maps, likely a reflection of increased tumor cellularity.

Most lesions show enhancement, which is often strong and homogenous (Fig-3). Marked edema and mass effect are uncommon. Hemorrhage and necrosis are rare [5].

Calcification is also rare, but may be seen post therapy [4, 5].

PCNSL in Immunocompromised Patients:

The mean age at presentation is 35 years. Multifocal lesions are more common, seen in about 30-80% of patients [1, 8].

Lesions most commonly involve the basal ganglia (Fig-4), followed by frontal, parietal and temporal lobes. Overall, the involvement of posterior fossa, corpus callosum and periventricular regions is more common. Most lesions are iso to hypo intense on T1W but may be hyper intense in up to 25% of cases due to hemorrhage [8]. Foci of bleeding may also be seen on GRE images (Fig-5).
The lesions show variable enhancement and non-enhancing lesions can also occur. Enhancement is typically irregular and peripheral and may be ring like in up to 75% of cases [1, 8] (Fig-6).

**Imaging findings suggestive of PCNSL:**

- Presence of perivascular spread is highly suggestive of PCNSL (Fig-7), with sarcoidosis representing the only other consideration [1, 2]. On microscopy, the lymphomatous cells may be seen to extend along the perivascular spaces (Fig-8) and infiltrate vessel walls.
- Similarly, bulky infiltration of corpus callosum (Fig-9,10,11) that is not accompanied by necrosis is suggestive of PCNSL, although it may also be seen in low grade gliomas [7].
- Patchy or diffuse coating of ventricles is also suggestive of the diagnosis (Fig-12) and is seen in about 38% of cases [3].

**Uncommon presentations of PCNSL:**

- Smaller lesions may sometimes mimic infarcts [5]. Clinically, however, the symptoms do not correspond to a stroke. In such cases, close follow up may be helpful (Fig-13-16).
- Uncommonly, PCNSL may present with diffuse infiltration of brain parenchyma [4, 5]. There is preservation of blood brain barrier and enhancement is therefore absent.
- PCNSL may present with a primarily leptomeningeal or dural involvement without any parenchymal lesions [1, 5].

**Secondary involvement in systemic lymphoma:**

Secondary intracerebral involvement may be seen in up to 10-15% cases of systemic lymphoma [9].

Intracranial spread from systemic disease (Fig-7) may involve the leptomeninges (with or without parenchymal lesions) or the dura (Fig-17, 18). It is exceptional for parenchymal lesions to occur without leptomeningeal spread [10].

Administration of intravenous contrast and post contrast FLAIR imaging improve detection of leptomeningeal disease which is rather poorly seen on CT and non contrast MR imaging [2, 9].
At times, progressive ventricular dilatation on serial scans may be the only telltale sign of leptomeningeal spread.

**Ocular involvement in lymphoma:**

In cases of PCNSL, this is usually due to extension of parenchymal disease to the eye. It has been reported in up to 25% of cases of PCNSL and is usually asymptomatic [1, 5].

On imaging, it may manifest as nodular enhancing lesion at the macula or as thickening of uvea. Primary intra ocular CNSL is otherwise very rare.

In cases of systemic lymphoma, ocular involvement may occur and have similar imaging findings (Fig-19,20).

**Imaging pitfalls in intracranial lymphoma:**

- Imaging after administration of steroids: These may be administered to treat raised intra cranial pressure. They decrease lesion enhancement on post contrast images. Additionally, they cause lymphocyte apoptosis and may reduce lesion bulk/ cause necrosis.
- Previous lumbar puncture may cause CSF leak and induce intra cranial hypo tension. In such cases, post contrast images may show dural thickening and enhancement and point towards a lymphomatous involvement. For this reason, lumbar puncture should be avoided before neuroimaging [1].

**Advanced imaging in intra cranial lymphoma:**

These may be used as problem solving tools to diagnose PCNSL.

- On perfusion imaging, PCNSLs demonstrate low CBV and a characteristic intensity time curve due to leakage of contrast in to interstitial spaces [1, 2].
- MR spectroscopy may reveal presence of elevated lipid peaks and high Cho/Cr ratios [11].
- Similarly, the fractional anisotropy values in PCNSL are significantly lower than in GBM and may aid in differentiating these tumors [1].
- PCNSL show higher metabolic activity on FDG PET (Fig-21) as compared to metastases/ high grade gliomas [12].
Images for this section:

**Fig. 0:** Axial T2W image reveals left peri ventricular mass which is iso intense to the grey matter. There is slight perilesional edema.

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Fig. 0: Same lesion as Fig-1.DW image shows restricted diffusion.

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**Fig. 0**: Same lesion as Fig-1. Post contrast coronal image reveals solid enhancement within the mass.

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Fig. 0: PCNSL in HIV. Axial T2W image reveals a heterogeneous lesion involving the right basal ganglia with moderate peri lesion edema and mass effect. Abnormal signal is also noted in the head of left caudate nucleus.

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**Fig. 0:** PCNSL in HIV, same patient as Fig-4. Axial GRE image reveals foci of blooming within the lesions, in keeping with blood degradation products.

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**Fig. 0:** PCNSL in HIV, same patient as Fig-4. Post contrast axial image reveals ring enhancement on right side without any significant enhancement on left side. PCNSL in HIV can present with both enhancing and non enhancing lesions with in the same patient.

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**Fig. 0:** T1W axial post contrast image reveals presence of subtle linear enhancement along the peri vascular spaces (black arrow) in right periventricular region. Also note enhancing subependymal nodules anterior to the large mass.

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Fig. 0: H&E (x 60) from a different patient reveals perivascular infiltration by atypical lymphoid cells (black arrows).

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Fig. 0: PCNSL involving corpus callosum. Axial T2W image shows bulky infiltration of corpus callosum with no evidence of any necrosis.

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**Fig. 0:** PCNSL involving corpus callosum, same patient as Fig-9. Axial DWI shows typical restricted diffusion.

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**Fig. 0:** PCNSL involving corpus callosum, same patient as Fig-9. Axial post contrast image reveals moderate enhancement within the lesion.

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Fig. 0: Axial post contrast CT image reveals presence of patchy subependymal enhancement involving bilateral lateral ventricles (black arrows). Associated involvement of choroids plexus is also noted bilaterally.

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Fig. 0: Bipsy proven lymphoma mimicking an infarct. Initial T2W image of a patient who presented with features of left sided weakness reveal a hyper intense lesion in right thalamus.

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**Fig. 0:** Bipsy proven lymphoma mimicking an infarct, same patient as Fig-13. Axial DW image at presentation reveals restricted diffusion within the lesion.

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Fig. 0: Bipsy proven lymphoma mimicking an infarct, same patient as Fig-13. Since the clinical findings were discordant a follow up MR was obtained after 3 weeks. T2W image from the second study reveals marked increase in size of the lesion with associated perilesional edema.

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**Fig. 0:** Bipsy proven lymphoma mimicking an infarct, same patient as Fig-13. Axial post contrast image from the second study reveals presence of rim enhancement within the lesion.

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**Fig. 0:** Axial T1W pre contrast image in a patient with CNS relapse of systemic lymphoma. There is presence of a hemorrhagic lesion involving the left temporal lobe. Altered signal is also noted in the right temporal lobe.

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**Fig. 0:** Same patient as Fig- 18. Axial post contrast image reveals enhancement with in the hemorrhagic lesion along with extensive leptomeningeal involvement on right side.

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**Fig. 0:** Intraocular relapse of systemic lymphoma. There is presence of a small plaque like lesion involving the region of macula in the left ocular globe on T1W precontrast image.

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**Fig. 0:** Intra ocular relapse of systemic lymphoma, same patient as Fig-20. The lesion shows prominent post contrast enhancement. Given the history and concurrent systemic relapse of lymphoma, a tissue diagnosis of the intra ocular lesion was not obtained.

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**Fig. 0:** Same patient as Fig-4. FDG PET CT shows increased uptake within the non necrotic right basal ganglia lesion (black arrowhead) and left caudate head.

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Lymphoma in CNS can be a great imitator. It's rising incidence and therapeutic implications makes awareness of its imaging manifestations all the more important.
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