Burkitt Lymphoma Presenting as Left Lateral Rectus Palsy

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Burkitt Lymphoma Presenting as Left Lateral Rectus Palsy

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Case:

- A 43 year old male approximately 2 weeks prior to initial hospital presentation had some left eye pain while shoveling snow. The following day he woke up with left eye pain and double vision. He was seen by his primary doctor who prescribed him eye drops to reduce the pain, but still had double vision. On physical exam he was found to have impaired left eye abduction and ptosis consistent with palsy of cranial nerves VI and III, respectively. He did not have other neurologic pertinent findings such as slurred speech, weakness, numbness, facial weakness. He was evaluated in the emergency department. Initially, his CT scan did not show any acute stroke. His first MRI showed no evidence of acute infarct but showed abnormal soft tissue fullness in the cavernous sinus, especially on the left with concern for lymphoma or leukemia with metastatic disease or sarcoidosis. Basic laboratories including CBC and CMP were within normal limits apart from a mildly elevated AST of 42.
- CT scan of chest, abdomen, and pelvis to uncover the source of cavernous sinus masses. Results showed gastric antral wall thickening (Fig 1), extensive peritoneal carcinomatosis, reticulonodular opacities within the lungs, a pancreatic tail mass, small bilateral renal lesions and upper abdominal lymphadenopathy. The patient was initially treated with palliative brain radiation until diagnosed with lymphoma.

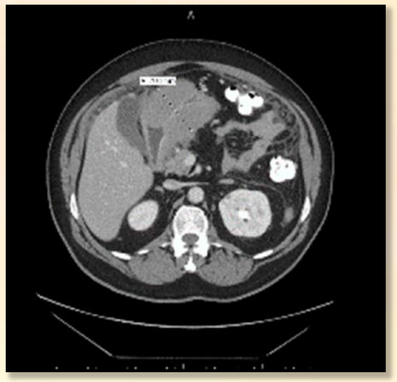


Figure 1. Gastric antral wall thickening on CT.

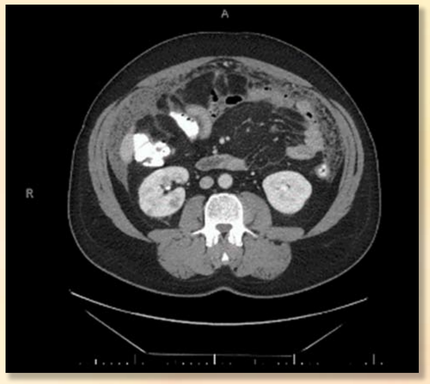


Figure 2. Peritoneal carcinomatosis on CT.

Table 1. Diferential Diagnosis of Visual Disturbance

Presentation	Differential
Unilateral Painful Vision Disturbance	• Corneal abrasion or infection • Acute angle-closure glaucoma • Iritis/uveitis • Optic neuritis • Endophthalmitis • Cavernous sinus thrombosis/mass
Diplopia	• Thyroid disease • Cranial Neuropathyy (CN III, IV, VI) • Neuromuscular disease (Myasthenia gravis) • Botulism, Miller-Fisher Syndrome • Encephalitis, Basilar meningitis • Brianstem Stroke

- EGD and biopsy of the gastric lesion (Fig 3) as well as bone marrow biopsy (Fig 4, 5) of the iliac spine. Ki-67 index was very high >90%, c-MYC rearrangement was present, and his disease was determined to be stage IVB. Immunohistochemical studies:

CD20: (+)	Ki-67: (+) in	BCL-2: (-/ dim focal+)
CD10: (+)	approx. 90%	BCL-1: (-)
BCL-6: (+)	of nuclei	TdT: (-)

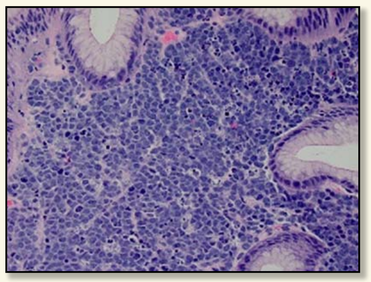


Figure 3. Gastric biopsy showing "starry sky" pattern.

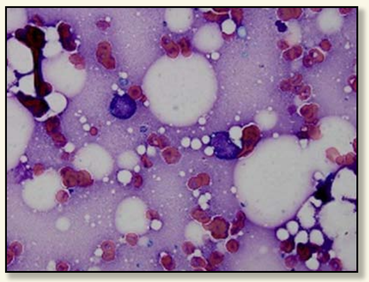


Figure 4. Bone marrow showing lymphocytic vacuolization.

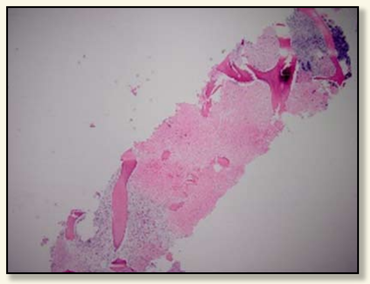


Figure 5. Bone marrow showing extensive necrosis.

Discussion:

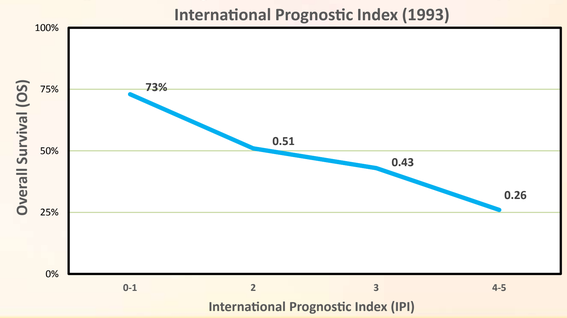
- Malignant metastasis to the central nervous system affects as many as 25% of patients with cancer.[1]
- Burkitt lymphoma (BL) is an uncommon[2] aggressive B-cell Lymphoma composed of rapidly proliferating B cells; it is considered the most highly aggressive NHL, with doubling times of 24 to 48 hours in some cases.[3]
- BL is subdivided into Endemic, Sporadic, and immunodeficiency-associated.
- Diagnosis of BL requires IHC staining for CD20+, CD10+ and TDT- with delineation from DLBCL by negativity for BCL-2.
- Translocation between MYC on chromosome 8 and one of three immmuoglobulin chain loci results in overexpression of the MYC protein causing deregulation of cellular growth.[3]
- Initial presentation of BL as cavernous sinus syndrome is uncommon.[4]
- Patient underwent treatment with HyperCVAD with intrathecal Methotrexate for CNS prophylaxis.
- The patient's prognosis was calculated using the International Prognostic Index (IPI) for aggressive lymphoma5. The IPI is a model for predicting outcome based on patient clinical characteristics prior to treatment. Patients are stratified according to risk with implications for treatment modalities. Those at high risk who are not effectively treated with current regimens may benefit from new clinical trial therapeutics. There are 5 adverse prognostic risk factors based on features of tumor growth potential, patient response to tumor, and ability to tolerate therapy: 1) age > 60 yo, 2) Ann Arbor Stage III/IV, 3) > 1 extranodal site, 4) Serum LDH > normal, 5) ECOG performance status >/= 2. (Score range 0 - 5).
- The patient's IPI : 2 (Stage IVB and >1 extranodal site) indicated a 5 year overall survival of 51% and complete response rate of 67%.

Table 2. Ann Arbor Staging System for Lymphomas[2]

Stage	Cotswold Modification of Arbor Classification
I	Involvement of a single LN region or lymphoid structure.
II	Involvement of two or more LN regions on the same side of the diaphragm (Mediastinum is considered a single site, but the hilar LNs are considered bilaterally); the number of atomic sites should be indicated by a subscript (e.g., II_3).
III	Involvement of LN regions on both sides of the diaphragm: III1 (with or without involvement of splenic hilar, celiac, or portal nodes) and III2 (with involvement of paraaortic, iliac, and mesenteric nodes.
IV	Involvement of one or more extranodal sites in addition to a site for which the designation E has been used.

Risk Group	IPI Score	% Patients	5-year OS	Complete Response Rate
Low	0-1	35%	73%	87%
Low-Intermed	2	27%	51%	67%
High-Intermed	3	22%	43%	55%
High	4-5	16%	26%	44%

Figure 6. Prognosis Variables for NHL Subtypes[5]



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