NHS England

Protocol for management of patients with suspected primary CNS lymphoma v1

West Midlands Cancer Alliance

Coversheet for Cancer Alliance Expert Advisory Group Agreed Documentation

This sheet is to accompany all documentation agreed by the West Midlands Cancer Alliance Expert Advisory Groups. This will assist the Clinical Network to endorse the documentation and request implementation.

EAG name	Brain and Central Nervous System	
Document Title	Protocol for management of patients with suspected primary CNS lymphoma	
Published date	April 2018	
Document Purpose	This guidance has been produced to support the management and follow-up of adult patients with suspected and proven primary CNS lymphoma tumours.	
Authors		
Review Date (must be within three years)	April 2021	
Approval Signatures:	**EAG Chair**  Date: 20 April 2018	**Network Clinical Director**  Date: 20 April 2018

PROTOCOL FOR MANAGEMENT OF PATIENTS WITH SUSPECTED PRIMARY CNS LYMPHOMA

Date Approved	**April 2018**
Date for Review	**April 2021**

DOCUMENT HISTORY

Version	Date	Summary
1	April 2018	Reviewed by Cancer Alliance Expert Advisory Group

Scope of the Guideline

This guidance has been produced to support the management and follow-up of adult patients with suspected and proven primary CNS lymphoma tumours.

2. Background

2.1 Primary CNS lymphoma is uncommon with an incidence of about 4.8 per million population per year and form about 5% of all brain tumours.

2.2 Focal neurological deficits are the commonest presenting symptom (70%), followed by raised intracranial pressure (33%) and seizures (14%).[i] Behavioural changes are common

2.3 Intra-ocular lymphoma can occur with CNS lymphoma, manifesting as a steroid resistant posterior uveitis.

2.4 The commonest site of the brain to be involved are the frontal lobe, corpus callosum, basal ganglia and periventricular white matter. They can be multi-focal.

2.5 Haemorrhage and calcification are very rare in lymphomas.

2.6 Diffuse large B-cell lymphoma comprises approximately 90% of cases with T-cell lymphoma about 5%. Low-grade B-cell lymphomas also occur (marginal zone B-cell lymphoma and lymphoplasmacytic lymphoma); this group often present as leptomeningeal or dural masses, mimicking meningiomas.

2.7 Primary CNS lymphoma can regress on steroids, but relapse within a short time of stopping steroids. The response to steroids is not diagnostic as it can occur with many neuro-inflammatory diseases and sarcoidosis.

2.8 Patients with primary CNS lymphoma should be screened for extracranial disease.

2.9 The British Neuro-oncology Society, in collaboration with the national Cancer Action Team, have produced guidelines for the management of primary CNS lymphoma, and this pathway document is based on those guidelines.

3. Guidelines

3.1 Patients with suspected lymphoma should be discussed in a neurosciences MDT prior to biopsy.

3.2 Steroids should not be given to patients with suspected cerebral lymphoma prior to biopsy.

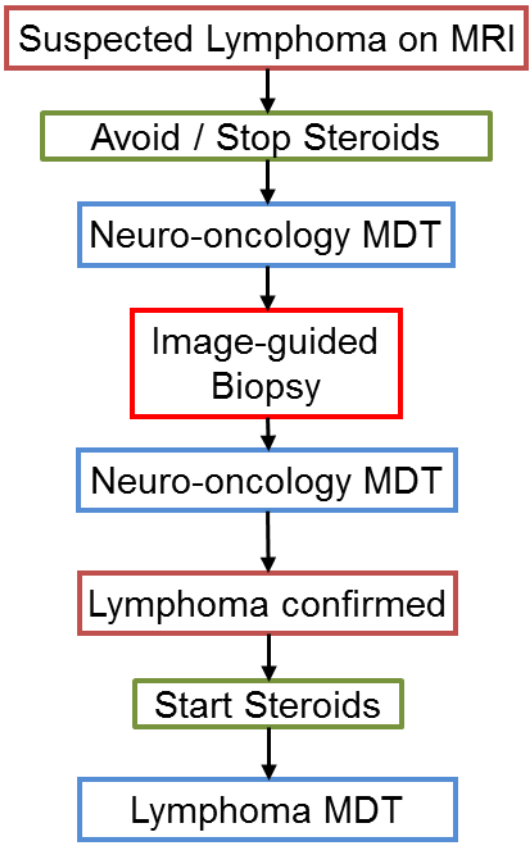
3.3 Biopsy should be performed by a core MDT member using appropriate image-guided surgery equipment.

3.4 Steroids can be commenced once a definitive histological diagnosis has been obtained.

3.5 Patients with confirmed cerebral lymphoma should be referred to the appropriate lymphoma MDT for further treatment. Further treatment will be according to the lymphoma MDT policies.

3.6 Patients who have had a suspected tumour regress on steroids should have their steroids stopped and should be rescanned at 3 months initially, unless symptoms recur earlier. Lesion regression on steroids should not be regarded as diagnostic of lymphoma.

4. Flowcharts



[i] Bataille *et al*. Primary intracerebral malignant lymphoma: A report of 248 cases. *Journal of Neurosurgery* 2000;**92**:261-6.