

## CHAPTER 19 PRIMARY CEREBRAL LYMPHOMA

### 19.1 Introduction

Primary cerebral lymphoma (PCL) is defined as a lymphoma confined to the central nervous system without evidence of systemic disease. It is an uncommon entity, but appears to be increasing in frequency in immuno-competent individuals.<sup>1</sup> No large randomised trials exist to dictate appropriate management and treatment of this condition. Recommendations are based on level III evidence. Objective responses to treatment are common and may be of long duration, but the disease frequently recurs, even many years after initial diagnosis and treatment.

The management of PCL requires referral to a specialised centre with multidisciplinary care.

### 19.2 Staging

There is no defined staging process. As a minimum, patients require a CT or MRI scan of the brain. Additional investigations may include: ocular examination, CSF examination and systemic staging including bone marrow biopsy and CT scan of chest and abdomen.<sup>2</sup> HIV testing is indicated in at risk groups.

### 19.3 General comments on treatment

No randomised studies have examined the most appropriate form of therapy for PCL and there is no standard therapy. However, most major centres use either chemotherapy alone or chemotherapy in combination with radiotherapy.

### 19.4 Surgery

All patients require a tissue diagnosis, but aggressive surgical resection does not result in a survival benefit and is not indicated.<sup>3</sup>

Guideline — Primary cerebral lymphoma — biopsy	Level of evidence	Refs
Patients with suspected primary cerebral lymphoma require biopsy only rather than resection.	III	3

### 19.5 Radiotherapy

Whole brain irradiation results in improved survival.<sup>1</sup> Dose escalation or non-standard fractionation provides no additional survival benefit.<sup>4</sup> Radiotherapy may be used as salvage treatment in patients who have previously been treated with chemotherapy.

### 19.6 Chemotherapy

Chemotherapy appears to provide significant improvements in survival above those achieved by irradiation alone. High response rates can be achieved with durable complete responses, but require agents that effectively cross the blood–brain barrier. Many strategies are effective in producing objective responses, including: single-agent high-dose methotrexate, chemotherapy with blood brain barrier disruption, combination intravenous and intrathecal therapy, and combination chemotherapy and radiotherapy.<sup>1,5–10</sup>

Guideline — Primary cerebral lymphoma — chemotherapy	Level of evidence	Refs
Patients with primary cerebral lymphoma may be treated with chemotherapy alone or chemotherapy in combination with radiotherapy.	III	1, 5–10

## 19.7 Toxicity

Both radiotherapy and chemotherapy are associated with cognitive deficits, particularly when intravenous and/or intrathecal Methotrexate is given following radiotherapy.<sup>8</sup> Single-agent high-dose methotrexate is well tolerated in elderly patients.<sup>7</sup>

## 19.8 References

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