Introduction

Primary orbital non-Hodgkin’s lymphoma (NHL) is a rare presentation of extranodal NHL accounting for less than 1% of cases [13]. A marked increase in the incidence has been observed over the last decade [35]. However, with a few exceptions, almost all reported studies have included a relatively small number of patients from one institution; therefore, a reasonably meaningful estimate of pretherapeutic or therapeutic prognostic factors has been difficult to establish [33]. Various subsytes within the orbit may be involved, including the conjunctiva, lacrimal apparatus, eyelids and musculature. Although many histologic subtypes of NHL may be encountered, the extranodal marginal zone lymphoma, mucosa associated lymphoid tissue (MALT) type is the most common subtype [21]. Several prognostic factors such as age, anatomical location of tumour, stage, histological subtype, grade, lactate dehydrogenase level, complete response to treatment, have been identified that could be used when considering the overall management of this uncommon site of primary lymphoma [11, 33]. Radiotherapy (RT) is the most effective treatment for local disease either as the sole treatment for low grade lymphoma or in combination with chemotherapy for intermediate and high grade lymphoma [15]. In the recent years, targeted therapy has become an important additional treatment modality for NHL and rituximab is the most commonly used [17].

Clinical characteristics

The presenting symptoms of primary orbital lymphoma (POL) vary according to the site of involvement. Common symptoms are diplopia, ptosis, pain, changes in visual
acuity, or restriction of eye movement; signs may include soft tissue swelling arising from the conjunctiva, eyelids or lacrimal gland or proptosis due to the orbital tumour [37]. With the exception of pain with associated bone erosion on radiological examination (most often observed in so-called high-grade lymphomas), there are few symptoms or signs that aid the clinical distinction between reactive lymphoid hyperplasia and malignant lymphoma [11]. Very few studies have systemically assessed the implications of these symptoms on outcome [6]. POL can be seen at all ages, but it is most commonly diagnosed in patients in the fifth to seventh decades of life [16]. POL tends to affect women more often than men, and occurs most frequently in the orbit and the conjunctiva [19]. Approximately 10–17% of cases demonstrate bilateral involvement, with the bilaterality occurring either simultaneously or subsequently [11]. The non-ophtalmic findings discovered on physical examination depend on the presence or absence of systemic malignant lymphoma.

**Diagnosis, histopathological classification and staging**

A detailed investigation as in other lymphoma sites is necessary, and the diagnosis of POL requires appropriate imaging studies. The CT scans and magnetic resonance imaging (MRI) are usually the primary diagnostic tools in the evaluation and both are of use to determine the size, the number and the degree of infiltration of these lesions. However, neither CT scans nor MRI can reliably differentiate lymphoid hyperplasia from malignant lymphoma [20, 40]. The final diagnosis is usually established through histopathological examination, which also helps with the histological classification of lymphoma. A thorough staging workup is necessary to identify those patients who may have additional foci of lymphoma. The staging of disease includes complete blood count, bone-marrow biopsy, immunophenotyping, CT scans of the neck, chest, abdomen and pelvis, and should now include positron emission tomography (PET) [5]. The role of PET was shown in detecting systemic disease compared to traditional total body scans [46]. A limitation of PET however is the low sensitivity in detecting orbital lesion, markedly lower than with traditional imaging, but this could be overcome by the advent of PET/CT, which combines the advantages of both techniques [41]. The majority of orbital lymphomas are B-cell NHL, which are predominantly extranodal marginal zone B-cell lymphomas [11], according to the Revised European American Lymphoma (REAL) classification [24] but they can also include diffuse large cell B-cell lymphomas and follicular lymphomas as well as less common B-cell lymphoma subtypes. Orbital lymphomas of non-B-cell type are rare in these sites [26], Hodgkin’s disease of the ocular adnexa is exceptionally rare, with only a few cases being reported in the literature [27]. POL is thought to be isolated to the orbit as the only extranodal site of involvement, thus by definition POL is stage I. Approximately 60–80% of POL are limited in their extension at the time of diagnosis and represent stage IE disease [2, 12, 42]. Bilateral involvement without systemic lymphoma is reported in about 10-17% of the cases but no consensus has been reached with regard to the classification of bilateral disease and depending on the author, it can be referred to as Stage I, II, IE, IIE or IIEE [33].

**Prognosis**

Various prognostic factors have been described as having an influence on the clinical course of the patients with orbital lymphoma. Major documented prognostic