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KEY POINTS

- Head and neck (HN) sarcoma and lymphoma share similarities in being uncommon mesenchymal tumors where the goal of treatment should emphasize methods of achieving disease control while maximizing preservation of cosmesis and function.
- Cross-sectional imaging is best undertaken in sarcomas before any biopsy or intervention to avoid compromising the planning of appropriate management; in lymphomas this is also needed for local radiotherapy (RT) but should be especially remembered if systemic treatment is undertaken initially to facilitate subsequent RT planning.
- Pathological grading provides an important prognostic measure and has been incorporated into the staging system of sarcomas.
- Clear surgical margin resection with or without RT is almost exclusively the mainstay of management of HN sarcoma. The indication for adjuvant systemic treatments is generally based on the experience of sarcomas in more common anatomic sites elsewhere.
- HN bone sarcomas differ from soft tissue lesions in that RT is much less established as a component of treatment, but chemotherapy is usually indicated in osteosarcoma and the very rarely seen HN Ewing’s tumor.
- In the combined RT and surgery approach to HN sarcoma, preoperative RT seems particularly suited, because of the smaller RT volumes and lower doses to critical anatomy where wide margins cannot be obtained.
- The management of non-Hodgkin’s lymphoma (NHL) in the HN is primarily influenced by histology and the Ann Arbor stage.
Optimal pathology interpretation of NHL involves immunophenotypic and sometimes molecular studies. Although surgical treatment for lymphoma is rarely required, open surgical approaches for tissue acquisition may be necessary.

- Stages I and II indolent lymphomas are treated with low to moderate dose involved-field RT with the expectation of a local control rate exceeding 95%, and long-term disease-free survival of 50–70%.
- Aggressive histology lymphomas presenting in early stage (I–II) is still associated with high risk for occult systemic disease and the use of anthracycline-based chemotherapy is required.

8.1 Introduction

Sarcoma and lymphoma are tumors of mesenchymal origin that comprise a contrasting subset of uncommon head and neck (HN) neoplasms. Lymphomas enjoy excellent response to chemotherapy and radiotherapy (RT) and almost never require surgery. In contrast, sarcomas almost always require surgery to achieve local control, and rarely develop regional metastases other than for some uncommon histological subtypes so that elective neck management is generally unnecessary.

Sarcomas comprise less than 1% of all HN cancers and their management follows principles extrapolated from other anatomic sites where the disease is more common (O’Sullivan et al. 2007). Surgery with or without RT is almost exclusively the mainstay of management. The indication for adjuvant systemic treatments relies for the most part on published meta-analyses for soft tissue sarcoma (STS) and some overviews for HN osteosarcoma.

HN lymphomas can present in lymph nodes or in extranodal organs. In virtually all cases a philosophy of functional preservation without resorting to ablative surgery should be followed. The need for chemotherapy and/or radiation therapy is not diminished following surgical excision of lymphoma. Advanced stages (stages III–IV) of lymphoma are treated primarily with chemotherapy and will not be further discussed. For stage I–II presentations in the HN area, the treatment approach involves radiation alone, or combined modality therapy (CMT) with chemotherapy followed by consolidation radiation, with curative intent. The description of management that follows will address two broad categories, specifically indolent lesions and the contrasting group with aggressive histology lymphoma.

While dramatically contrasting in terms of management approaches, both HN sarcoma and lymphoma share similarities in that the goal of treatment should emphasize methods of achieving disease control while maximizing preservation of cosmesis and function. This philosophy will be maintained in the discussion that follows, although no formal quality of life or functional assessment studies specific to these HN tumors are available. Some illustrative cases will complement the discussion to demonstrate anatomic challenges that can be met with individualized approaches to the choice of RT and surgical techniques.

8.2 Evaluation and Diagnosis

8.2.1 Sarcoma

Local management of STS and bone sarcoma starts with imaging and physical examination to determine whether soft tissue lesions originate superficial or deep to the muscle investing fascia and whether bone tumors are intramedullary or have invaded into the extra-osseous compartment. Cross-sectional imaging is best undertaken before any biopsy or intervention to avoid compromising the planning of appropriate management since even needle tracts are potential sites of tumor contamination. Ideally the surgeon and radiation oncologist should be involved in planning the optimal biopsy route and be aware of potential contamination. Computerized tomography of the chest is required, excepting very low grade lesions where a plain chest radiograph may be sufficient because of the low risk of distant metastases.

The prototypical STS has been malignant fibrous histiocytoma but additional unique predispositions in the HN include angiosarcoma of the scalp and facial regions, hemangiopericytoma of the sinonasal region, dermatofibrosarcoma protruberans of the dermal regions of the low neck and supraclavicular area and rhabomyosarcoma which preferentially