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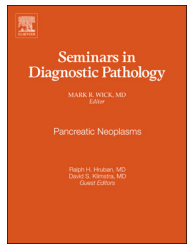

Hematolymphoid lesions of the sinonasal tract

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ABSTRACT

Various hematolymphoid lesions involve the sinonasal tract, including aggressive B, T, and NK-cell neoplasms; myeloid sarcoma; low-grade lymphomas; indolent T-lymphoblastic proliferations; and Rosai–Dorfman disease. Differentiating aggressive lymphomas from non-hematopoietic neoplasms such as poorly differentiated squamous cell carcinoma, olfactory neuroblastoma, or sinonasal undifferentiated carcinoma may pose diagnostic challenges. In addition, the necrosis, vascular damage, and inflammatory infiltrates that are associated with some hematolymphoid disorders can result in misdiagnosis as infectious, autoimmune, or inflammatory conditions. Here, we review hematolymphoid disorders involving the sinonasal tract including their key clinical and histopathologic features.

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Introduction

Hematolymphoid disorders that involve the nasal cavity and paranasal sinuses range from aggressive and rapidly fatal neoplasms to relatively indolent disease. Lymphomas are the most common hematolymphoid disorder of the sinonasal tract, with sinonasal disease comprising approximately 1% of all lymphomas in Western countries.[1,2] The majority of sinonasal lymphomas diagnosed in Western countries are B-cell neoplasms, and the most common lymphoma involving the sinonasal area is diffuse large B-cell lymphoma (DLBCL).[1–3] Low-grade B-cell neoplasms can also involve the sinonasal region and often present with symptoms of congestion in adults or as difficulty breathing in children.[1,2,4,5] T-cell lymphomas are far less frequent than B-cell lymphomas, although one of the best known sinonasal lymphomas is extranodal natural killer (NK)/T-cell lymphoma, nasal type, with its aggressive nature reflected in its former moniker "lethal midline granuloma."[5,7] The diversity of hematolymphoid disorders that may involve the sinonasal tract combined with the rarity of some of these entities and potentially limited material in biopsy specimens can result in diagnostic challenges.

Aggressive B-cell neoplasms

Diffuse large B-cell lymphoma

Diffuse large B-cell, not otherwise specified (DLBCL, NOS, or DLBCL), is the most common type of sinonasal lymphoma seen in Western populations, and it represents 33–58% of all sinonasal non-Hodgkin lymphomas and 42–94% of B-cell lymphomas involving the sinonasal tract.[1–5] In a large series described by Mian et al.,[6] DLBCL affecting the sinonasal tract had a median age of onset of 61 years (range: 21–91) and a similar incidence in male and female patients. Unlike T-cell and NK/T-cell lymphomas, which often arise in the nasal cavity itself, DLBCLs tend to arise in the paranasal sinuses.[3,5] Common symptoms at presentation include nasal congestion,

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soft tissue swelling, localizing pain, rhinorrhea, and epiphora.[3] Most patients with DLCBL involving the head and neck are treated with chemotherapy and/or radiation therapy. Patients in the Mian, et al.[6] series demonstrated a similar complete response rate, 5-year overall survival, and 5-year event free survival when compared to patients with DLBCL involving other sites of the head and neck.

DLBCL is a heterogeneous disease, but most present as sheets of large, atypical lymphocytes. The chromatin can range from fine to vesicular and the nucleoli may be prominent or inconspicuous. Apoptotic bodies and mitotic figures are often readily appreciated. DLBCLs characteristically express pan B-cell markers including CD19, CD20, CD22, and CD79a, and surface and/or cytoplasmic immunoglobulin can be demonstrated in most cases.[7] The proliferative index as measured by Ki-67 immunostaining is high; it is often at least 50% and may exceed 90%. A subset of sinonasal DLBCLs are associated with Epstein–Barr virus (EBV) (Fig. 1), and rare cases of ALK-positive DLBCL involving the sinonasal tract have also been reported.[5,8,9]

Burkitt lymphoma

Burkitt lymphoma (BL) is the second most common aggressive B-cell neoplasm affecting the sinonasal region after DLBCL, and accounted for 3–11% of the B-cell lymphomas that arose in the sinonasal tract in several large series.[1,2,4,5] Variants of BL include endemic, sporadic, and immunodeficiency-associated BL, any which may present with sinonasal involvement.[9] Endemic BL is limited to equatorial Africa, but sporadic BL is seen worldwide and most commonly presents in children and young adults.[7] Immunodeficiency-associated BL is predominantly seen in patients with HIV infection, and sinonasal BL has been reported in this context, including at least two reports of HIV+ adults who presented with symptoms of chronic sinusitis.[10,11] BL is an aggressive neoplasm, but cure rates may reach as high as 60–90%.[7]

Morphologically, BL is characterized by sheets of medium-sized atypical lymphocytes with a high nuclear to cytoplasmic ratio, round nuclei with finely clumped chromatin, and multiple paracentrically located basophilic nucleoli. On touch preparations or smears, the neoplastic cells have deeply basophilic cytoplasm and often contain characteristic lipid vacuoles. There are numerous apoptotic bodies, and the scattered tingible body macrophages that engulf the apoptotic debris give rise to the characteristic “starry sky” appearance of BL. Tumor cells are positive for CD19, CD20, CD10, Bcl-6, CD45, and surface light chain, but they are typically negative for Bcl-2. Nearly 100% of nuclei label with Ki-67, reflecting the high proliferation index. Virtually all BLs demonstrate a translocation joining *MYC* to the immunoglobulin heavy or light chains.[7] Of note, *MYC* rearrangements are not specific for BL and can also be seen in DLBCL and plasmablastic lymphoma.[7,12]

B-lymphoblastic leukemia/lymphoma

The differential diagnosis of BL includes B-lymphoblastic leukemia/lymphoma, which may also involve the nasal

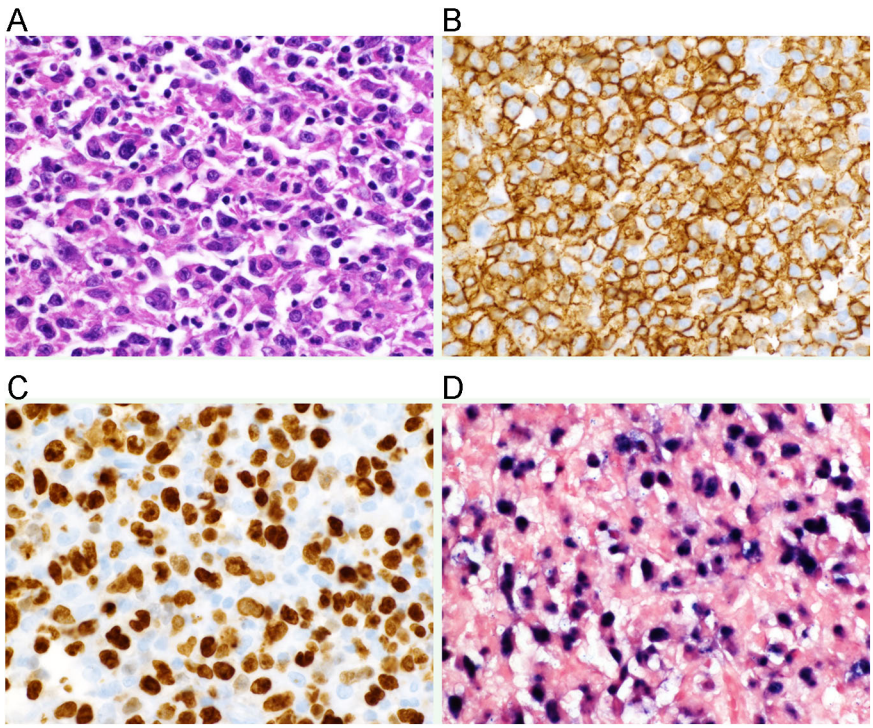

Fig. 1 – EBV-positive diffuse large B-cell lymphoma of the elderly. A nasopharyngeal biopsy of a mass in a 60-year-old woman shows sheets of large atypical lymphoid cells with abundant cytoplasm and irregular nuclear borders with variably prominent nucleoli (A, H&E). The tumor cells express CD20 (B) and have a high Ki-67 proliferation index (C). An *in situ* hybridization for EBV-encoded small RNA (EBER) is positive in the neoplastic cells (D). All images, 400 ×.

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