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Schnoz and Proboscis- Sinonasal B Cell Lymphoma

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Sinonasal tumours incriminating nasal cavity or paranasal sinuses are extremely exceptional and constitute a minute component (~ 3%) of neoplasms arising within upper respiratory tract. Malignant tumours of sinonasal tract predominantly occur within maxillary sinus followed in frequency by ethmoid sinus, frontal sinus or sphenoid sinus.

Discernment of a unilateral nasal mass mandates cogent tissue sampling wherein possible emergence of lymphoma within nasal cavity or paranasal sinuses requires elimination. Preliminary diagnosis and pertinent staging appear imperative for efficacious therapy.

Sinonasal B cell lymphoma is an exceptional, malignant, non Hodgkin's lymphoma incriminating nasal cavity, paranasal sinuses, sinonasal region or circumscribing soft tissues. Sinonasal lymphoma may manifest as a singular, isolated neoplasm or appear in conjunction with systemic disease [1,2].

Incidence of sinonasal lymphoma is enhanced within Asian population whereas the lymphoma is uncommon within Caucasians. Lymphoma occurring within Asians may arise within nasal cavity. Sinonasal B cell lymphoma configures a significant component of extranodal lymphoma, following gastrointestinal lymphoma in frequency.

Sinonasal B cell lymphoma is accompanied by favourable prognostic outcomes, in contrast to extra-nodal natural killer cell lymphoma (ENKL) which exhibits a rapidly progressive clinical course and disease associated mortality. Appropriate histological discernment with pertinent immunohistochemistry is beneficial [1,2].

Of rhinologic origin, majority (~90%) of sinonasal B cell lymphomas are preponderantly comprised of diffuse large B cell lymphoma (DLBCL). Additionally, primary lymphomas as high grade B cell lymphoma (HGBCL), high grade B cell lymphoma double hit (HGBCLDH), high grade B cell lymphoma not otherwise specified (NOS), mantle cell lymphoma, low grade B cell lymphoma, plasmablastic lymphoma (PBL), lymphoplasmacytic lymphoma, plasmacytoma or Burkitt's lymphoma (BL) may be delineated [1,2].

Sinonasal B cell lymphoma commonly implicates nasal cavity or paranasal sinuses as maxillary sinus, ethmoid sinus, frontal sinus, sphenoid sinus or antronasal region. Sinonasal B cell lymphoma develops within an anatomic space and expands toward paranasal sinus, nasal cavity or nasopharynx. Incriminated ethmoid sinus is associated with destruction of orbital plate.

A male predominance is observed with male to female proportion of ~1.5:1 [1,2].

Generally, middle aged adults between 5th decade to 6th decade are implicated although the lymphoma may appear between 21 years to 88 years. Average age of disease representation is 49 years whereas median age of disease discernment is 49 ± 12.2 years [1,2].

Majority of instances accompanied by delayed tumour detection generally demonstrate initial and prominent sinonasal symptoms. Besides, preliminary neoplasms manifest as localized disease.

Factors contributing to occurrence of sinonasal B cell non Hodgkin's lymphoma are comprised of autoimmune disease, infection with human immune deficiency virus (HIV) or autoimmune deficiency syndrome (AIDS), infection with human T lymphotropic virus (HTLV) and exposure to immunosuppressant agents or pesticides. Besides, malignant sinonasal B cell lymphoma may be discerned within immunocompetent subjects [1,2].

Sinonasal B cell non Hodgkin's lymphoma frequently demonstrates genetic mutations within p53, K-RAS, C-KIT, β-catenin or BAK genes. Thus, it is posited that precise genetic mutations may configure the trigger inducing sinonasal B cell lymphoma [1,2].

Preliminary disease is usually devoid of pertinent clinical symptoms. Following initial non specific symptoms, cogent clinical manifestations may occur as nasal congestion, nasal obstruction, facial swelling, pain, epistaxis, rhinorrhoea, non healing ulcer, headache, septal perforation, bone destruction, cervical lymphadenopathy, proptosis, loss of vision, B clinical symptoms as fever, night sweats, 10% loss of body weight or rhinosinusitis [1,2].

Clinical representation of sinonasal lymphoma is contingent to histological subtype wherein low grade lymphoma exhibits a sinonasal tumefaction along with obstructive symptoms and regional lymphadenopathy [1,2].

High grade lymphoma manifests with aggressive symptoms as non healing ulcer, cranial nerve injury, facial swelling, epistaxis, pain, bony destruction or proptosis.

Regional lymph node metastasis or systemic, B clinical symptoms may be observed upon initial disease representation [1,2].

Diffuse large B cell lymphoma (DLBCL) may occur as unilateral lesion confined to nasal cavity or maxillary sinus. Subsequently, the lymphoma demonstrates widespread dissemination into maxillary sinus, ethmoid sinus, sphenoid sinus or orbit.

Extra-nasal neoplastic dissemination is infrequently encountered within regional lymph nodes, cutaneous surfaces and testes. Primary sinonasal diffuse large B cell lymphoma may exhibit contiguous dissemination into regional cervical lymph nodes [1,2].

Disseminated or preceding or secondary sinonasal diffuse large B cell lymphoma may manifest singularly as extra-nodal lymphoma, singular nodal lymphoma or an admixture of extra-nodal and nodal lymphoma. Upon microscopy, the commonly discerned diffuse large B cell lymphoma exemplifies diffuse infiltration of scattered, enlarged or intermediate, atypical B lymphoid cells. Neoplastic cells are incorporated with vesicular chromatin and prominent nucleoli. Enlarged cells with enlarged nuclei demonstrate nuclear magnitude ≥ histiocytic nuclei or > 2 lymphocytic nuclei. Diffuse large B cell lymphoma exhibits morphologic variants denominated as immunoblastic variant, centroblastic variant or anaplastic variant. Cellular component may simulate anaplastic large cell lymphoma or Reed-Sternberg cells [1,2].

TNM staging of sinonasal B cell lymphoma [2,3]:

- **Primary tumour**
    - T1: Tumour confined to nasal cavity
    - T2: Tumour extension into maxillary sinus, anterior ethmoid sinus or hard palate

- T3: Tumour extension into posterior ethmoid sinus, sphenoidal sinus, orbit, superior alveolar bone, cheeks or superior buccinators space
- T4: Tumour incrimination of inferior alveolar bone, inferior buccinators space, infratemporal fossa, nasopharynx or cranial fossa.

- **Regional lymph nodes**
  - N0: No regional lymph node involvement
  - N1: Unilateral regional lymph node involvement
  - N2: Bilateral regional lymph node involvement.
- **Distant metastasis**
  - M0: Distant metastasis absent
  - M1: Distant metastasis present.

Pertinent immuno-phenotype and cytogenetics may be evaluated wherein sinonasal B cell lymphoma is immune reactive to CD3, CD20, BCL2, BCL6, cMYC along with Epstein Barr virus encoded RNAs (EBER) [3,4].

Lymphomas with genomic rearrangement of MYC may be analysed for chromosomal rearrangements of BCL2 and BCL6 [3,4].

Cogent disease discernment can be obtained with comprehensive clinical history and physical examination. Assessment of factors such as age at initial diagnosis, gender, pertinent clinical symptoms, occurrence of systemic symptoms, serum lactate dehydrogenase (LDH) levels, localization of tumour reoccurrence and dissemination, adopted therapeutic strategies and response to treatment along with cause of mortality appear advantageous [3,4].

International Prognostic Index (IPI) pertaining to age of incriminated subject is beneficially assessed as per World Health Organization (WHO) or Eastern Cooperative Oncology Group (ECOG) performance score.

Serum LDH at initial disease detection may be above upper limit of normal [3,4].

Peripheral blood involvement may occur with variants such as mantle cell lymphoma or low grade B cell (LGBCL) lymphoma. Computerized tomography (CT) or magnetic resonance imaging (MRI) can be adopted for localized tumour staging and detection of distant metastasis [3,4].

Contrast enhanced computerized tomography (CT) and magnetic resonance imaging (MRI) are beneficially employed to assess extent of tumour, bone destruction, precise tumour staging and pertinent site for tissue sampling [3,4].

Cogent chemotherapeutic regimen are comprised of cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) or a CHOP-like regimen may be adopted as initial treatment. Besides, additional chemotherapeutic regimen or immunotherapy may be employed. Consolidative radiotherapy may be advantageously utilized for treating diverse subtypes of low stage disease [3,4].

Central nervous system (CNS) prophylaxis may be achieved with enhanced doses of methotrexate, cytarabine, methotrexate- cytarabine combination or triple CNS therapy with intrathecal methotrexate, cytarabine and hydrocortisone [3,4].

5 year proportionate overall survival for the predominant primary sinonasal diffuse large B cell lymphoma is ~ 56%. Persistent cervical lymphadenopathy mandates a thorough examination of nasopharyngeal system [3,4].

Extra-nodal tumour relapse of sinonasal lymphoma beyond gastrointestinal tract may ensue within larynx, cutaneous surfaces, hepatic or renal parenchyma, uvula, breast, lacrimal gland, testis or prostate gland [3,4].

Prognostic outcomes are contingent to stage and subtype of disease, quantifiable sites of extra-nodal tumour dissemination, infiltration of central nervous system and general health of incriminated individual.

Inferior prognostic outcomes are obtained with

- Age of incriminated subject > 60 years
- ECOG performance status
- Involvement of regional cervical lymph nodes.
- High grade lymphoma or reoccurring, disseminated disease [3,4].

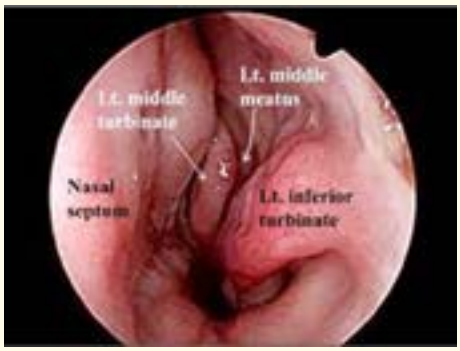

Figure 1: *Sinonasal B cell lymphoma confined to frontal sinus delineating a tumour mass devoid of superimposed ulceration or haemorrhage [5].*

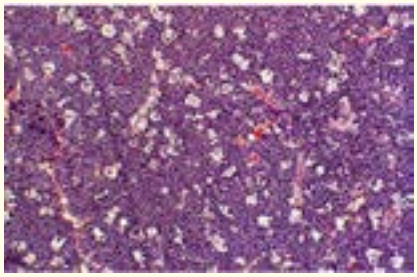

Figure 2: *Sinonasal B cell lymphoma demonstrating dense, diffuse infiltration of atypical lymphocytes with moderate cytoplasm, vesicular nuclei and conspicuous nucleoli, intermingled with congested capillaries and delicate fibro-connective tissue [6].*

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5. Image 1 Courtesy: Science direct.
6. Image 2 Courtesy: OOOOjournal.net.

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