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Case report

Lymphoma Maxilla mimicking orbital cellulitis; case report and review of Literature

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Abstract

Background: Sinonasal lymphomas are the commonest nonepithelial malignancies of the nasal cavity and paranasal sinuses. They may spread from their site of origin in nasal cavity and invade adjacent structures including orbits leading to varying presentation and a diagnostic dilemma. **Case:** A-77- year old Nepalese male presented with swelling of left eye since 1 month and pain in the same eye since 2 days. **Observation:** Examination revealed axial proptosis, swelling of lids, restricted extraocular movement, diffuse conjunctival and ciliary congestion with chemosis. Visual acuity was grossly decreased. The case was diagnosed as left orbital cellulitis and treated with broad spectrum intravenous antibiotics and oral steriods. There was no response despite 3 days of antibiotics and steroids therefore a computed tomography was planned which showed features of Sinonasal malignancy invading the orbit. Repeated biopsy was suggestive of malignant small round cell tumour. In the view of unclear preoperative biopsy indicating further details on the histologic type of tumor and extensive erosion of maxilla with complete loss of vision in the eye the patient was planned for Total Maxillectomy with Orbital extenteration of the left orbit. Post- operative histopathological and Immunohistochemical report were suggestive of High grade Lymphoma. The patient was advised for Adjuvant Chemotherapy and Radiotherapy, which the patient refused due to financial constraints. He is on occasional follow up since the last 6 months. **Conclusions:** The diagnosis of maxillary sinus lymphoma needs to be borne in mind when a clinician encounters a case of Orbital Cellulitis. It may be difficult to diagnose clinically and require radio-pathological correlation.

Keywords: Lymphoma, Maxillary sinus, orbital cellulitis

Introduction

Lymphoma is a general term for malignant tumors arising from the lymphocyte. These tumors are relatively uncommon, representing less than 1% of all head and neck malignancies (Keyser et al, 2000). They have been traditionally divided into Hodgkin's disease and non-Hodgkin's disease-the former characterized by the presence of the Reed-Sternberg cells and the later by its absence. NHL can further be

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subdivided into T-cell, B-cell, or natural killer cell lymphomas depending on the lymphocytic cell of origin. Lymphomas may originate from a lymph node or lymphatic tissue outside the lymph node (extranodal lymphomas). The extranodal lymphoma as mainly involve 4 sites: Waldeyer's ring, the nasal sinuses and fossae, the oral cavity and the salivary glands (Picarda et al, 2015).

Lombard et al (2015) state that the main diagnostic problem is the lack of specific clinical signs, often leading to late consultation and thus diagnosis at an advanced stage. A lymphoma in the maxillary sinus may have myriad presentations. Clinical presentation largely consists of local signs such as unilateral nasal obstruction, purulent and/or blood- streaked rhinorrhea, recurrent epistaxis or chronic sinusitis (Mestiri et al 2008; Lachiver et al 1995). General signs, such as fever, weight-loss or night sweats, are rare and concern advanced stages. Lachiver et al(1995) state that signs of regional extension (jaw swelling or ophthalmic, auditory, pharyngeal or neurological disorder) may be the presenting symptoms in late stages.

Sinonasal lymphomas are the commonest nonepithelial malignancies of the nasal cavity and paranasal sinuses. In Asian population, over 90% of these sinonasal lymphomas are T cell lymphomas. (Kamath et al 2006). Sinonasal lymphoma may spread from their site of origin and involve the surrounding structure including orbit. Clinical presentations of orbital lymphoid neoplasms are most commonly characterized by slow, painless onset of swelling of the eyelid, palpable mass and proptosis. Other common symptoms may include diplopia, diminished visual acquity, pain, ptosis and restricted eye movement (Gupta et al 2010). In 22.5% of patients, Sino-nasal lymphoma and orbital lymphoma may co exist and such patients need collaborative management efforts by Opthalmologists, Otolaryngologist and Oncologist. However to date in our knowledge there are no current literatures in which orbital cellulitis as the presenting feature of Sino-nasal lymphoma has been reported. We hereby report a case which presented as orbital cellulitis to the Opthalmology Department of our Tertiary care centre and on further work up turned out to be B cell Lymphoma of the maxilla.

Case Report

A 77 years Nepalese male presented in the Opthalmology Out patient department with swelling of left eye (Fig. 1 and 2) since 1 month, which increased since last 7 days and gradually spread to the left cheek. The swelling was also associated with pain over the left eye since 2 day. On examination the best corrected visual acuity (BCVA) was 6/12 in the contralateral right eye with pseudophakia and normal fundus findings. The BCVA was ½ /60 with no improvement with Pin hole in the involved left eye. There was axial proptosis with diffuse swelling of upper lid, lower lid and left cheek region with extraocular movement restriction in all cardinal positions of gaze. Conjunctiva showed diffuse and ciliary congestion with chemosis but cornea and anterior segment examinations were normal. Pupillary reaction, fundus examination, Optic Nerve function test and IOP in both eyes were also normal.

The case was diagnosed as left orbital cellulitis and was admitted and treated with broad-spectrum intravenous antibiotics and oral steriods. As there was no response despite 3 days of antibiotics and steroids, a computed tomography of orbit was planned which showed permeative pattern of bone destruction involving inferior, anterior and lateral wall of the left maxillary sinus, zygomatic process and zygomatic bone with wide zone of transition with large homogenously enhancing soft tissue component suggestive of primary malignant bone tumour (Fig.3 and 4).

On the basis of CT scan findings the opinion

of the ENT colleagues was sought. There was no telecanthus or hypertelorism but proptosis of left eye with edematous upper and lower eyelids with crusting and pus discharge from lower eyelid. Oral examination revealed a firm, nontender bulge in left side of hard palate. On anterior rhinoscopy, a proliferative growth was seen in left middle meatus. On examination of the neck trachea was central and no lymph nodes were palpable. Endoscopic biopsy from growth in left middle meatus showed nonspecific rhinosinusitis. As endoscopic biopsy was inconclusive therefore Incisional biopsy left maxilla by sublabial approach was planned. It revealed tumour cells with round, hyperchromatic nuclei, irregular nuclear membrane, inconspicuous nucleoli and scanty cytoplasm which were predominantly singly scattered and occasionally arranged in cords. Fair number of individual cell necrosis, apoptotic cell bodies, necrotic debri and thick walled blood vessels lined by plump endothelial cells were also seen, suggesting malignant small round cell tumour (Fig.5). In the view of unclear preoperative biopsy indicating further details on the histologic type of tumor and extensive erosion of maxilla with complete loss of vision in the eye the patient underwent Total Maxillectomy by the combined ENT and Orbital extenteration of the left orbit by the Ophthalmology team.

The Post- operative period was uneventful and the patient was discharged on the 7th Post-operative day. Immunohistochemical report showed that it stained LCA, CD20,CD79a (Fig.6) but the tumour did not express CD3, NSE, Desmin, MyogeninSmooth muscle actin, CK, CD99, anti MPO and CD56 suggestive of High grade Lymphoma of the B phenotype. The patient was advised for Adjuvant Chemotherapy and Radiotherapy, which the patient refused due to financial constraints. He is on occasional follow up since the last 6 months ((Fig.7 and 8).

Fig. 1 and 2 : Orbital cellulitis at presentation

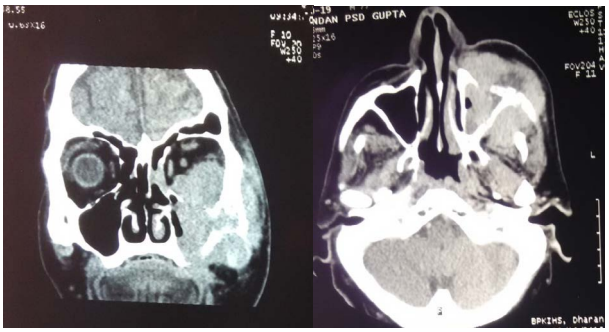

Fig. 3 and 4: CT scan showing extensive tumor destruction of maxilla extending to the orbit

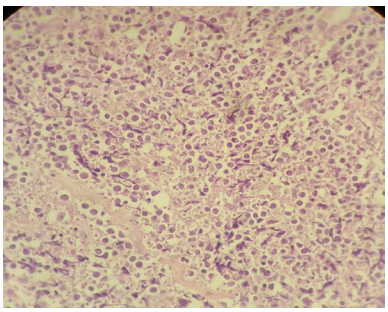

Fig.5: Photomicrograph revealing singly scattered small round cells having irregular hyperchromatic nucleus. Individual cell necrosis and apoptotic cell bodies are also seen (40X, H&E)

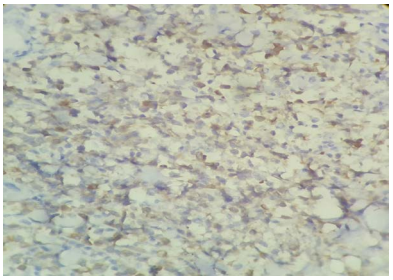

Fig. 6 : Section revealing tumor cells positive for CD 20 (40x, IHC)

Fig. 7 and 8 : Post operative Follow up.

Discussion

Malignant lymphoma is the second-most common malignancy in the head and neck region, followed by squamous cell carcinoma (Epstein et al, 2001). Two major classes of lymphoma are Hodgkin disease and non-Hodgkin lymphoma. Hodgkin disease typically arises in the lymph nodes and has a predilection for cervical and mediastinal nodes whereas NHL often presents as extranodal disease (Triantafillidou et al, 2012). Otter et al (1989) state that approximately 40% of NHL arises from extranodal sites outside the lymphoid system and the most common site of extra-nodal NHL in the head and neck region is Waldeyer's ring.

Margo et al (1998) and Moslehi et al (2006) state that over the last 4 decades, there has been a reported increase in the number of systemic and ocular NHL cases, which may largely be a result of advances in medical diagnostic testing and changes in classification of lymphomas. In addition, NHL has been linked to environmental factors, such as pesticides, wood preservatives, and solvent exposure; immune disorders, such as occurs with patients on immune-suppressants or those with a transforming virus such as human immunodeficiency virus or Epstein Barr virus; patients with connective tissue disorders, such as Sjogrens, lupus, or rheumatoid arthritis; and infectious diseases, such as chlamydia or hepatitis C (Margo et al, 1998; Moslehi et al 2006; Engels, 2007).

ENHL type of lymphoma affects patients with a median age of 68 years, with peak incidence in the 6th and 7th decades, although all ages may be concerned. Patients are more often male (Picarda et al, 2015).This is similar to the reported case.

Lombard et al (2015) in the largest published series of 22 sinonasal lymphoma so far found that the presenting symptoms were poorly specific, with classic sinonasal functional signs: unilateral nasal obstruction, mucopurulent rhinorrhea, epistaxis and anosmia. They also found a considerable number of patients (16 out of 22) who showed non-rhinologic signs: diplopia (27%), exophthalmus (18%), endo-oral swelling (18%) or cranial nerve palsy (9%) and cervical adenopathy(1 patient). A tumor in the maxillary sinus may produce symptoms depending upon the direction of growth. Medial spread from the maxillary sinus to the nasal cavity may produce nasal obstruction or epistaxis, posterior spread to the pterygopalatine fossa may invade the pterygoids leading to trismus, anterolateral spread may lead to cheek swelling, inferior spread causes dental pain, loosening of molars or endo- oral swelling and predominantly superior growth lead to orbital symptoms. There are numerous reports of maxillary lymphomas mimicking dental conditions and presenting to the dental surgeons as odontogenic infection, osteomyelitis or a soft tissues swelling in the oral cavity (Van der Waal et al, 2005; Kolokotronis et al, 2005), periodontal abscess (Yepes et al, 2006), mucous retention pseudocyst on panoramic radiography or as ulcerated growth of upper alveolus (Adwani et al, 2013). In our case the patient did not have any rhinological symptoms but predominantly orbital because the tumor had grown superiorly eroding the roof of maxilla and involving the orbit whereas the medial wall of the maxilla, the nasal cavity and the turbinates are all free of tumor as also evident on the CT-scan (Fig.3 and 4).

On the contrary it is not uncommon for both

orbital and extra-orbital tumors to present as orbital cellulitis. There are reports of Retinoblastoma (Mullaney et al,1998), Adenocarcinoma oesophagus with secondary orbital metastasis (Kean et al,2000) and Acute Myelogenous Leukemia (Bagheri et al, 2013) presenting as orbital cellulitis but however to our knowledge this is the first report where a lymphoma in the maxillary sinus was seen to mimic orbital cellulitis. Sharma et al (2011) reported two cases of patients with a previous history of endoscopic nasal surgery presenting with progressive facial swelling mimicking cellulitis, followed by ulceration. After repeated inconclusive biopsies, immunohistochemical studies were undertaken, enabling the diagnosis of peripheral natural killer T cell lymphoma.

Clinical examination generally finds an anterior ulceronecrotic lesion, bleeding on contact and filling the nasal cavity, usually located on the lateral wall. Lymph-node involvement varies from series to series and never exceeds 25% of cases (Mestiri et al,2008). This lack of lymph node involvement could have been one of the reasons for the initial mislead in our case. The other cause for the delay in diagnosis in our case could have been that the lesion was located mainly in the anterolateral wall and roof of the maxilla. Direct tumour invasion in to the orbit could have been responsible for the inflammation and orbital cellulitis whereas the obsecure anterolateral maxillary wall location of the tumour may have made the deeper endoscopic biopsy from the lesion difficult giving a picture of nonspecific rhinosinusitis on preoperative biopsy which was overcome later with a biopsy via the sublabial approach. Though routine biopsies via the sublabial approach for maxillary sinus tumors are contraindicated, this may be one of the scenarios where they may seem useful. Morphologic assessment is the same as for other lymphomas, comprising contrast-enhanced CT-scan, completed by PET-CT.

NHL can be managed by chemotherapy, radiotherapy and surgery in various combinations. In our case the patient had undergone Total Maxillectomy with Orbital extenteration because of extensive erosion of maxilla with complete loss of vision in the eye and the preoperative biopsy indicating tumor lacked further detail as to histologic type; lymphoma was diagnosed only on definitive pathology examination and immunohistochemistry.

Most NHLs in the head and neck regions are malignancies of the B-lymphocytes, and diffuse large B-cell lymphoma is the most common type followed in frequency by mucosa-associated lymphoid tissue (MALT) lymphoma (Kemp et al, 2008). High grade Lymphoma of the B phenotype was also seen in our case.

Conclusion

Periorbital malignancies masquerading as orbital disease are common. ENT and Dental and sometime Neurosurgical consultation always advised as per presence of local signs. CT scan is a useful adjunctive diagnostic tool whose importance cannot be minimized even in nonmalignant cases- not only to rule out presence of orbital abscess in case of orbital cellulitis with vision involvement but also for decision regarding surgical intervention. Though rare maxillary lymphoma has to be borne in mind in a patient presenting with orbital cellulitis.

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