• Letter to the Editor •

Primary sino-orbital peripheral T-cell lymphoma presenting as unilateral periorbital swelling: a case report

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Dear Editor,

My name is Ahmad Al Omari and I am currently working as an otorhinolaryngology assistant professor at Jordan University of Science and Technology. I am writing this letter to present a case of a primary sino-orbital peripheral T-cell lymphoma that presented as unilateral periorbital swelling.

Primary sino-orbital lymphomas are among the lymphoproliferative lesions that affect the nasal cavity, the para-nasal sinuses, and the orbit. Sino-nasal and orbital lymphomas may co-exist; however, the vast majority is lymphomas of B-cell lineage which are usually associated with the orbit. We report a 57-year-old man who presented with a progressive unilateral periorbital swelling with subsequent rapidly progressive ophthalmoplegia due to a primary sino-orbital peripheral T-cell lymphoma. To the best of our knowledge, this is the first case of primary sino-orbital peripheral T-cell lymphoma to be reported in the Middle East.

Nasal-type extranodal natural killer (NK)/T-cell lymphoma (NKTL) accounts for about 1.4% of all non-Hodgkin's lymphomas. The nose is the prototypic and most common site of involvement, but identical tumors can involve the skin (5.4%), gastrointestinal tract (3.6%), soft tissue (3.8%), and testis (1.8%)[1]. NKTL is prevalent in Asians and South American populations[2].

The most common presentations of sino-nasal lymphomas are epistaxis, nasal obstruction, and nasal swelling while proptosis or hard palate perforation is less commonly seen[3]. On the other hand, the most common presenting symptoms for orbital lymphomas are eyelid swelling followed by palpable eyelid mass, diplopia or blurred vision, proptosis, pain, and lid erythema[4].

According to Declaration of Helsinki, a written voluntary informed consent was obtained from the patient for publication of this case report and accompanying images.

CASE PRESENTATION

A 57-year-old diabetic man presented with periorbital swelling, proptosis, nasal discharge, fever, malaise, and headache for a couple of weeks. The patient had a previous history of functional endoscopic sinus surgery (FESS) 2y before this presentation. That surgery was performed because of chronic rhinosinusitis, and during the procedure, a post nasal polypoidal tissue was found incidentally and biopsied but showed no malignancy.

Ophthalmic examination showed right eye proptosis of 26 mm as recorded using a Hertel exophthalmometer. The measurement for the left eye was 18 mm. The extraocular motility (EOM) of the right eye was initially limited to abduction of -2, with normal EOM of the left eye. Regarding visual acuity, the initial record was best corrected vision of 0.7 and 1.0 (Figure 1).

Ear, nose and throat (ENT) evaluation showed a yellowish nasal discharge in the right nostril, with significant hypertrophy of the inferior turbinates. No lymphadenopathy was noted. A culture was obtained which was later shown to be positive for methicillin-resistant staphylococcus aureus (MRSA). Computed tomography (CT) of the orbit and sinuses showed bilateral micro-erosions of both lamina papyracea, particularly on the affected side (Figure 2).

In addition, there was diffusely enhanced soft tissue swelling in the preseptal region of the involved eye with no intra-conal extension. The sinuses showed a pattern indicative of pan-sinusitis and all of the radiological findings were suggestive of periorbital cellulitis. Because the patient initially refused admission, he was treated empirically on an outpatient basis as a case of acute bacterial sinusitis and orbital cellulitis. Two days later, he was admitted for intra venous antibiotic treatment of MRSA with piperacillin-tazobactam (4.5 mg *q6h*) and vancomycin (1 g *bid*) according to sensitivity.

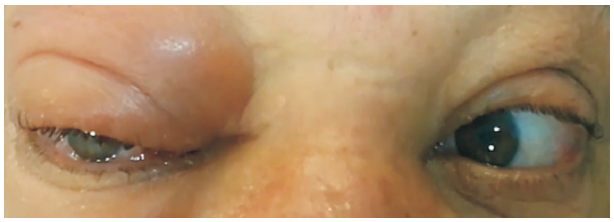

Figure 1 Right peri-orbital swelling with complete right eye EOM limitaion with gaze to the right side.

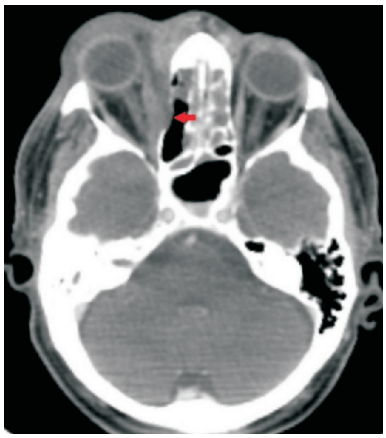

Figure 2 Orbital CT scan showing right sided proptosis as well as micro-erosions in both lamina papyreceae with diffusely enhancing soft tissue more prominent on the right.

Over 2 days, both EOM and visual acuity of the right eye deteriorated, accompanied by severe pain. As a result, he underwent endoscopic debridement. Intraoperatively, evidence of the previous endoscopic sinus surgery was noted; the turbinates in the nasal cavities were excessively hypertrophied obscuring the landmarks in both cavities (Figure 3).

Nasal biopsies were taken from the inferior turbinates. Pus was drained from the maxillary sinus. Anterior and posterior ethmoidectomy were revised during which multiple biopsies were taken from polypoidal tissue of the ethmoid sinus. A window was created in the right lamina papyracea to relieve the intra-orbital pressure and pus drainage was noted during this procedure.

After the surgery, the patient continued on intra-venous antibiotics but without significant improvement of his symptoms. Due to the slow progression of improvement, MRI of the paranasal sinuses with intravenous (IV). contrast was carried out. This enhanced the appearance of soft tissue in the right preseptal region along the lamina papyracea extending into the supraorbital region and involving the right medial and superior rectus muscles (Figure 4). No intracranial extension or skull base involvement was noted.

Histopathology findings were consistent with peripheral T-cell lymphoma and showed complete effacement of the architecture by typical medium to large sized lymphocytes, some with abundant clear cytoplasm and forming vague modularity as well as extensive necrosis (Figure 5A). The

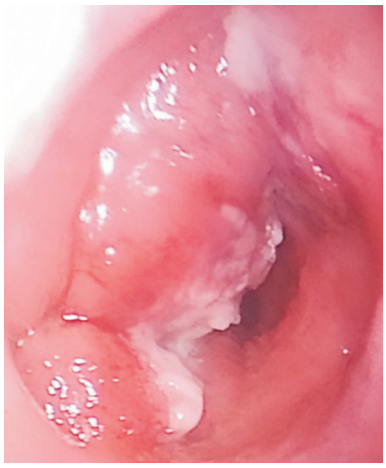

Figure 3 The inferior turbinate on the right nasal cavity side After application of 1:100 000 adrenaline-soaked neuropatties for 10min, and the right lateral nasal wall distorted anatomy.

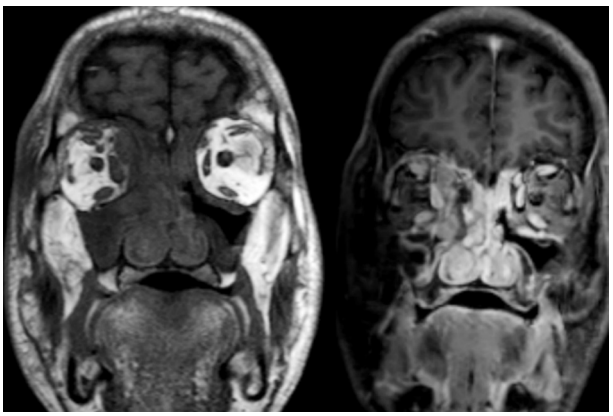

Figure 4 Paranasal sinuses and orbit T1-MRI showing difusely enhancing soft tissue involving the right preseptal region extending into the supraorbital region.

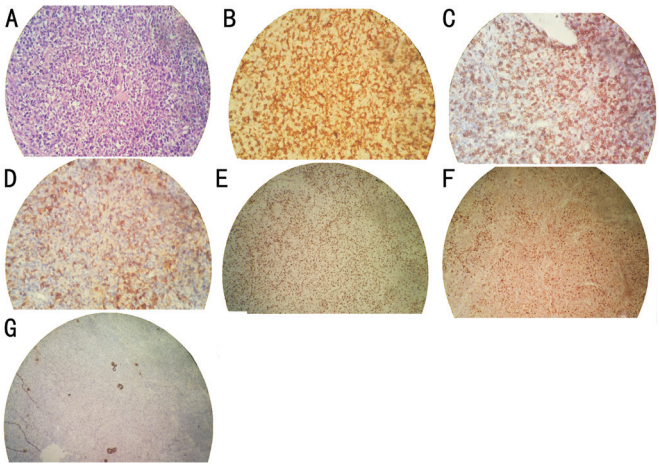

Figure 5 Histopathological Findings A: Complete effacement of the architecture by typical medium to large sized lymphocyte, some with abundant clear cytoplasm and forming vague modularity as well as extensive necrosis; B: CD3 T-cell marker; C: CD4; D: CD8; E: High Ki-67 proliferative index; F: Background showed few cells reactive to CD20; G: CK negative with internal control positivity (glands).

tumor cells were immunoreactive for CD3 (Figure 5B), LCA, CD4 (Figure 5C) and CD8 (Figure 5D) immunostains with a high

Ki-67 proliferative index (Figure 5E), and negative for pan-CK (Figure 5G), ALK, CD56, CD10, bcl-6, MUM-1, CD1a, TdT and CD34 immunostains. The background showed few cells reactive to CD20, PAX-5, and CD5 immunostains (Figure 5F). Epstein-Barr virus (EBV)-specific antibody serology testing was done and showed high viral capsid antigen (VCA) and Epstein-Barr virus nuclear antigen (EBNA) IgG antibodies. The oncology team was consulted and a metastatic work-up was performed consisting of neck, chest, abdomen and pelvis CT with IV and oral contrasts, lumbar puncture, and bone marrow biopsy; however, this showed no evidence of extra-sino-orbital tumor. The patient was started on chemotherapy [cyclophosphamide, hydroxydaunorubicin (adriamycin), vincristine (Oncovin), and prednisone (CHOP)], but his symptoms showed no improvement, so he is now receiving local radiotherapy.

DISCUSSION

Primary sino-orbital lymphomas are part of the lymphoproliferative lesions that affect the nasal cavity, the paranasal sinuses and the orbit. In nearly 22.5%, sino-nasal and orbital lymphomas may co-exist; however, lymphomas of B-cell lineage are more common and are more likely to be associated with symptoms related to the eyes (P<0.0005) and to have extension to the orbit (P<0.01) than lymphomas of T- or NK-cell lineage[5].

Sino-orbital N/K T-cell lymphoma is a rare disease, often with a fatal outcome. To the best of our knowledge, only a few cases have been reported in the literature. In one of the largest case series studies, a series 15 patients with nasal type extranodal NKTL was reported over 20y, with only four cases reported to have an orbital extension[6]. The non-specific symptoms of clinical presentation, such as nasal obstruction and nasal discharge in addition to the periorbital swelling, can be misleading and mistaken for chronic sinusitis with periorbital cellulitis[7].

Under the World Health Organization Classification, NKTL is the most common EBV+ form of lymphoma derived from NK cells and/or cytotoxic T-lymphocytes that typically involves the nasal cavity and paranasal sinuses[8]. It has been proposed that orbital involvement is usually a result of extension or invasion from the nasal cavity and paranasal sinuses with or without bony erosion[9].

For stage I/II nasal lymphoma, the best result can be achieved with a combination of chemotherapy and radiotherapy. For stage III/IV nasal lymphoma and non-nasal lymphomas, chemotherapy is the mainstay of treatment[10]; however, NKTL is highly aggressive, and can present initially with orbital and adnexal symptoms and has a poor prognosis once disseminated despite radiotherapy and aggressive chemotherapy[11].

In conclusion, an antibiotic non-responsive sino-orbital infection should be surgically debrided and studied histopathologically. Malignancies such as sino-nasal T-cell lymphoma can present as orbital swelling with nasal obstruction and can also have a rapidly progressive course.

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Conflicts of Interest: Al Omari A, None; **Al Zoubi F,** None; **Alsalem MM,** None; **Al-Salem KM,** None; **Mohidat H,** None.

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