CASE REPORT

Rapidly enlarging primary cutaneous anaplastic large T-cell lymphoma of the eyelid: A case report

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Abstract

Non-Hodgkin CD30+ T-cell lymphoproliferative disease encompasses three subgroups, one of which is primary cutaneous anaplastic large T-cell lymphoma (PC-ALCL). It is distinguished by the presence of giant anaplastic cells with pleomorphism and widespread CD30 positivity. The case of a 58-year-old female is presented who reported to the Eye Department of Jinnah Post-Graduate Medical Centre (JPMC) on 18th January, 2023 with a three-month history of rapidly enlarging, non-tender, ulcerated lesion on the lower eyelid of the right eye. The histopathology report identified the lesion as CD30+ primary cutaneous anaplastic large cell lymphoma. Computed tomography (CT) scanning of the orbit revealed no extension, and positron emission tomography-computed tomography (PET-CT) showed no systemic involvement. The patient underwent surgical excision without adjuvant chemo-radiation and has remained in clinical remission for five months.

Keywords: Eyelid Neoplasm, Anaplastic Large Cell Lymphoma, Eye Cancer, Eyelid Disease.

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Introduction

Primary cutaneous anaplastic large T-cell lymphoma (PC-ALCL) is one of the three subtypes of non-Hodgkin CD30+ T-cell lymphoproliferative disorders. It is characterised by the presence of large anaplastic cells exhibiting pleomorphic and diffuse CD30 positivity. Typically, PC-ALCL presents as a solitary erythematous ulcerated nodule,[1] while approximately 20% of patients may exhibit multifocal cutaneous involvement.[2] The other subtypes include lymphomatoid papulosis (LyP) and systemic anaplastic large-cell lymphoma (ALCL).[3]

PC-ALCL is a rare disease, with an overall incidence of 0.12 cases per 1,000,000 individuals (age-adjusted to the 2000 US standard population); approximately 658 cases have been reported in the literature from 1973 to 2016.[4] According to the current guidelines, the recommended first-line treatments include surgical excision and radiotherapy for solitary lesions. However, there have been reported cases where regression of solitary PC-ALCL was achieved with low-dose oral methotrexate.[5] In case of multifocal lesions or systemic involvement, adjuvant therapies such as chemotherapeutic agents are necessary in conjunction with the standard treatment.[6]

With the current treatment approach, the 5-year survival rate of PC-ALCL is between 80% to 90%.[1] However, due to its clinical presentation and rarity, PC-ALCL is often misdiagnosed for other, more commonly encountered tumours in outpatient settings. This case report aims to contribute to the existing scientific literature and expand our knowledge of PC-ALCL to improve diagnosis and management strategies.

Case Report

A 58-year-old female reported to the Eye Department of Jinnah Post-Graduate Medical Centre (JPMC), Karachi on 18th January, 2023. She presented with a three-month history of rapidly enlarging, non-tender ulcerated lesion on the lower eyelid of the right eye as shown in (Figure a). Prior to her subsequent presentation at JPMC, she had received initial treatment for presumed dacryocystitis in November 2022 at Noorani Eye Clinic Korangi, Karachi, a small medical centre near her area of residence which proved to be unsuccessful. Physical examination revealed a firm oval shaped lesion situated on the lower eyelid near the medial canthus of the right eye, measuring 17.0 x 12.0 mm in size. The lesion was non-tender, nodular and ulcerated, with

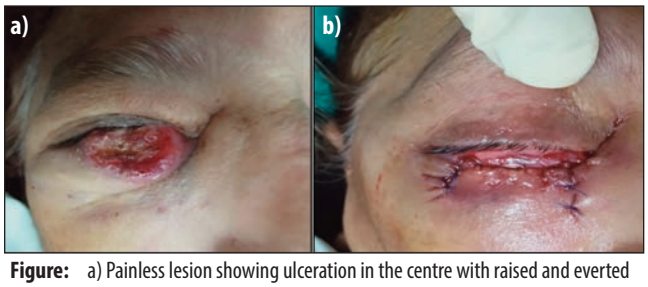

Figure: a) Painless lesion showing ulceration in the centre with raised and everted margins measuring to about 17.0 x 12.0 mm in size. b) After surgical excision and lid reconstruction with the Hughes flap.

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yellow and red coloured crusting in the centre and shiny, erythematous, everted margins. The surrounding skin appeared slightly erythematous. An incisional biopsy was obtained and sent for histopathological investigation. Concurrently, a range of diagnostic tests were ordered including PET-CT, contrast CT-Scan of the orbit, HBsAg, Anti-HCV, Bone Scan, and abdominal ultrasound to facilitate further evaluation. The histopathology report revealed diffuse sheets of enlarged cells with hyperchromatic nuclei, scant cytoplasm and scattered mitoses, including atypical ones. Immuno-histochemical staining was positive for CD3, CD30, CD45 (LCA) and Ki-67, with a high proliferative index of approximately 80%. PET-CT results indicated no systemic involvement, while orbital CT scan demonstrated a lobulated soft tissue density mass with heterogeneous post-contrast enhancement along the lower eyelid on the right side, exhibiting an exophytic component and ulcerated margins measuring up to 1.0 x 2.4 cm in size. This resulted in a 1.3 cm craniocaudal focal skin bulge. No calcifications were observed extending to the pre-septal region, and no retro-orbital extension was noted. HbsAg and anti-HCV were negative and the remainder of the investigations were within normal ranges. Following multidisciplinary discussion, wide surgical excision of the lesion and lid reconstruction with the Hughes flap was performed as shown in (Figure b), followed by the opening of the Hughes flap after six weeks. Chemoradiation was not advised as the lesional margins were found to be tumour-free. Since then, the patient has been monitored routinely and has remained in clinical remission for five months.

Discussion

PC-ALCL is among the three subtypes of non-Hodgkin CD30+ T cell lymphoproliferative disorders, which can present with similar phenotypic characteristics to other eyelid lesions.[7] Its subtypes, lymphomatoid papulosis (LyP) and systemic anaplastic large-cell lymphoma (ALCL) share phenotypic and histological similarities with PC-ALCL but can also be differentiated based on their clinical presentation.[3] LyP is a relatively benign subtype with a 4% chance of transforming into a malignant lymphoma.[2] It can be distinguished from PC-ALCL by the presence of multiple papulonodular lesions that follow a waxing and waning course.[8] Similarly, systemic ALCL with secondary cutaneous involvement is considered as the aggressive subtype with a less favourable prognosis and requires adjuvant therapies in addition to the standard protocol.[8] Although, histologically it is similar to PC-ALCL; however, the distinction is made based on clinical presentation. PC-ALCL lesions tend to occur in the periocular region, without systemic involvement in older patients (average age 61 years). Whereas, systemic ALCL commonly occurs on the lower extremities, with systemic manifestations, relatively in the younger patients (average age 24 years).[9]

Current guidelines recommend a CT or PET-CT imaging to rule out secondary cutaneous involvement by aggressive subtypes.[10] The patient's radiological investigation showed no systemic involvement; therefore, the surgical excision was suggested as the treatment of choice.

Other treatment modalities including chemotherapeutic agents such as cyclophosphamide, vincristine, doxorubicin, prednisone (CHOP) and Brentuximab-Vedotin (anti CD30 antibody) which are reserved for cases with extracutaneous involvement.[11] Further, the patient was referred to the oncology and radiology departments, but chemoradiation was not advised due to the limited nature of her lesion, with no systemic involvement.[12] Since then, the patient has been attending regular follow-ups and has been in clinical remission for five months.

Conclusion

To summarize, our patient, a 58-year-old female, was diagnosed with primary cutaneous anaplastic large cell lymphoma (PC-ALCL) after presenting with an ulcerated lesion on the lower eyelid. Histopathology report confirmed the case of PC-ALCL and imaging studies showed no systemic involvement. The lesion was treated with surgical excision only ensuring tumour free margins. The patient responded well to the treatment and has been in clinical remission after five months of follow-up.

Consent: Consent of the patient was obtained prior the writing and composing of this case report.

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Author Contribution:

VK: Concept, data acquisition, analysis, interpretation and final approval.
NA: Concept, data acquisition, analysis, interpretation, revision and final approval.
TF, NAK: Drafting, data analysis, interpretation and revision.