

Case Report

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Plasmacytoma of the Eyelid in an HIV-Positive Patient

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ABSTRACT

Extramedullary (extraosseous) plasmacytoma represents a rare disease that accounts for less 5% of plasma cell neoplasms. It commonly occurs in the upper respiratory tract, with 80% of cases involving the paranasal sinuses, nasopharynx, or nasal cavity. Plasmacytomas of the eyelid are very rare with only ten cases having been reported in the literature so far. We present the clinical and histopathologic findings from a case of plasmacytoma of the eyelid and discuss its histopathologic differential diagnoses. To our knowledge, this is the first case of plasmacytoma of the eyelid in an HIV-positive patient in the English literature.

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Introduction

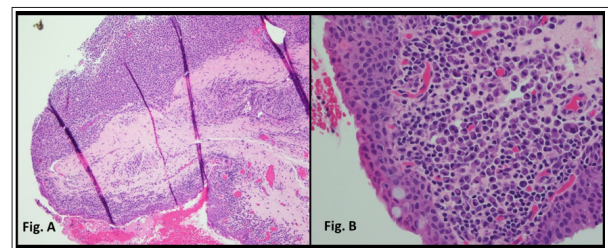
Extramedullary plasmacytomas are localized plasma cell tumors that arise in tissues other than bone marrow. They account for 4% of all plasma cell tumors with 75% to 80% of them occurring in the submucosa of the upper respiratory tract including the nasal cavity, paranasal sinuses, oropharynx, and larynx. They may also occur at other sites including lymph nodes, parotid, thyroid, GI tract, central nervous system, breast, testis, and other organs. Plasmacytomas of the eyelid are very rare with only 10 cases reported in the literature so far. Here, we present a case of plasmacytoma of the eyelid and discuss the histopathologic differential diagnoses [1-7].

Case Presentation

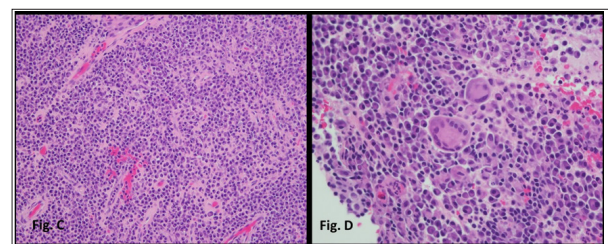
A 47-year-old HIV-positive male presented with a 1.3 x 1 cm left eyelid mass that was clinically diagnosed as chalazion. Histologic examination shows diffuse proliferation of mature plasma cells underneath the palpebral conjunctiva with areas of ulceration and few histiocytic giant cells. (Figs. A - D).

Kappa and lambda stains by in situ hybridization show two geographically distinct plasma cell populations. The superficial population of plasma cells is kappa-restricted, and hence is monoclonal (Fig. E); whereas the deeper population shows mainly lambda-positive plasma cells with fewer admixed kappa-positive cells. (Figs. F) Overall, there is significant kappa predominance with a ratio of approximately 5-10 to 1. PCR shows a clonal rearrangement of the immunoglobulin heavy chain gene.

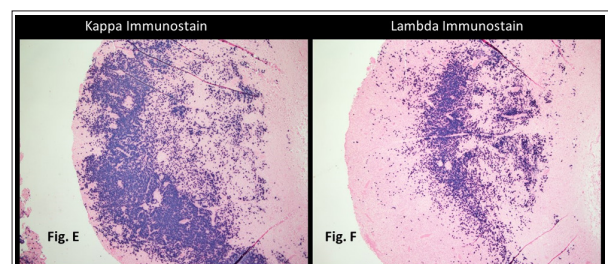
These findings are diagnostic of kappa-restricted plasmacytoma, located mainly in the superficial part of the specimen. However, given the presence of more lambda-positive cells in the deeper population, a biclonal process cannot be ruled out.



Figures A & B: Sheets of plasma cells underneath the conjunctival epithelium with goblet cells



Figures C & D: Medium and high-power views showing a diffuse proliferation of mature-appearing plasma cells with scattered histiocytic giant cells



Figures E & F: Kappa and Lambda Immunostains show two geographically distinct plasma cell populations. The picture on the left (Fig. E) shows that the superficial population of plasma cells is kappa-restricted (and hence is monoclonal), whereas the deeper population (Fig. F) shows mainly lambda-positive plasma

cells with admixture of kappa-positive cells.

Discussion

There are only few reports of extramedullary plasmacytoma in HIV-positive patients published in the literature, including those involving the kidney, testis, adrenal, and oral cavity [8-12]. Hematolymphoid tumors are a subset of HIV-related malignancies which are acquired immunodeficiency syndrome (AIDS)-defining malignancies. The major cause for the increased risk of development of these malignancies in HIV-positive patients compared to non-HIV infected individuals is immunosuppression. HIV infection has also been associated with increased incidence of unusual clinical presentation, and aggressive clinical behavior of extramedullary plasmacytoma cases. To the best of our knowledge, the present case is the first case of plasmacytoma of the eyelid associated with HIV infection [9]. Plasma cell tumors in HIV positive patients often has plasmablastic morphology. [13,14]. However, the current case does not show this morphology.

Extramedullary plasmacytomas present as a localized mass lesion. The median age at presentation is 55 years and two thirds of patients are males. Consistent with these clinical features, this 47 years old male patient presented with a left eyelid mass lesion that was clinically diagnosed as chalazion [15]. Histologically, it showed reactive histiocytic giant cells which could have been easily misinterpreted as part of a chalazion. However, the presence of numerous plasma cells lead to the consideration of plasma cell dyscrasia and the assessment for the clonality of the cells. Staining for kappa and lambda light chains showed kappa-restricted plasmacytoma. This case highlights the importance of staining for light chain restriction whenever one encounter diffuse plasma cells in eyelid specimens, more so in HIV patients even in the presence of some reactive histiocytic giant cells. It also highlights the need for careful evaluation of the kappa and lambda stains in the presence of significant number of both lambda and kappa positive plasma cells.

The other histologic differential diagnoses in this case are lymphoplasmacytic and extranodal marginal-zone lymphomas. Lymphoplasmacytic lymphomas consist of a mixture of clonally related plasma cells and lymphocytes usually with lymphocytes predominating. The absence of a significant lymphocyte component in the present case makes the diagnosis of lymphoplasmacytic lymphoma unlikely [15].

Extra-nodal marginal-zone lymphoma with plasma cell differentiation may mimic a plasmacytoma. However, the absence of the typical lymphoepithelial lesions, centrocyte-like cells with clear cytoplasm, or an admixture of cell types makes that diagnosis unlikely in this case.

The typical treatment of extraosseous plasmacytoma is local eradication by surgical excision and radiotherapy [2, 15]. This patient will need long-term follow-up as the possibility of local recurrence or spread to regional lymph nodes occurs in about 25% of extramedullary plasmacytomas and 10 – 30% of them progress to full blown multiple myeloma. Approximately 70% of patients with solitary extramedullary plasmacytoma remain disease-free at 10 years. [3, 14, 16, 17].

Conclusion

This case highlights the importance of staining for light chain restriction whenever one encounters diffuse plasma cells in eyelid specimens even in the presence of some reactive histiocytic giant cells. It also highlights the need for careful evaluation of the kappa and lambda stains in the presence of significant number of both

lambda and kappa positive plasma cells.

Disclosures

The authors do not have any potential conflicts of interest.

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