Case Report

Extramedullary plasmacytoma of the orbit: A rare case

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ABSTRACT

Solitary orbital plasma cell neoplasm in the absence of multiple myeloma is very rare. We present here a case report of extramedullary plasmacytoma of the right orbital soft tissue in a 50-year-old Indian woman whose thorough diagnostic workup was negative for multiple myeloma. The patient presented with gradually increasing proptosis, swelling, and visual disturbances of the right eye for past one and half years. Computed tomography (CT) scan of the orbit revealed lobulated, mildly enhancing soft tissue lesion in the supero-lateral aspect of right orbit without intracranial extension. Pathological report revealed a plasma cell neoplasm and immunohistochemistry was positive for CD138, CD20, CD 79a, PAX-5. Complete skeletal survey and bone marrow examination report were normal. So, normal skeletal survey, absence of bone marrow involvement, absence of Bence-Jones protein, no anemia, no hypercalcemia, no renal disease, normal level of immunoglobulins all exclude the diagnosis of multiple myeloma. Since the visual acuity of the patient is well-preserved, so we have planned to treat her with local radiotherapy alone with 50 Gy over 5 weeks.

Key words: Immunohistochemistry, orbit, plasmacytoma, rare

INTRODUCTION

Plasma cell neoplasms represent autonomous proliferations of plasma cells and can be manifest as systemic disease, namely, multiple myeloma and its variants (such as indolent myeloma, smoldering myeloma, osteosclerotic myeloma, plasma cell leukemia and non-secretory myeloma) or localized disease represented as solitary plasmacytoma of bone (SMB) or extramedullary plasmacytoma (EMP). Involvement of the orbit can occur in either of the two forms. Orbital involvement in multiple myeloma is very rare with less than 50 cases reported in the literature.[1,2] Most of the EMPs are located in nasal cavity and para-nasal sinuses.[3-5] A plasmacytoma involving the skull base or orbit in the absence of multiple myeloma is very rare. The nasopharynx is the skull base site most frequently involved, representing 18% of all head and neck cases. Sphenoid, clivus, and petrosal apex presentations are far less common.[6]

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However, an orbital plasma cell neoplasm in clear absence of multiple myeloma (as diagnosed by hematological, biochemical, and radiological investigations) is extremely rare.[7]

We present here a case report of orbital plasma cell neoplasm or EMP of the right orbital soft tissue in a 50-year-old Indian woman whose thorough diagnostic workup was negative for multiple myeloma.

CASE REPORT

A 50-year-old female was referred from The Regional Institute of Ophthalmology, Kolkata with redness and swelling of the right eye for past one and half years and progressive proptosis for past 6 months with moderate visual disturbances. Physical examination showed proptosis, chemosis, limited ocular movement, and swelling of the right eye. The visual acuity was 6/30 of the right eye. Computed tomography (CT) scan of the orbit revealed lobulated, mildly enhancing soft-tissue lesion at the supero-lateral aspect of right orbit. The tumor had no intracranial extension. Contrast CT brain was normal except senile atrophy.

Incisional biopsy was taken from the orbital soft tissue swelling, and the histopathology revealed fibrocollagenous

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tissue densely infiltrated by sheets of immature plasma cells, possibly of plasma cell neoplasm [Figure 1]. The slide was reviewed, and that revealed the same diagnosis of plasma cell neoplasm. The morphologic features were consistent with extramedullary plasmacytoma of soft tissue (of orbit) according to World Health Organization (WHO) classification. Immunohistochemical staining revealed that majority of the plasma cells expressed CD138, CD20, CD 79a, PAX-5. Kappa light chain was expressed, and lambda light chain and cytokeratin were negative.

Since the visual acuity of the patient is well-preserved, so we have planned to treat her with local radiotherapy alone with 50 Gy over 5 weeks.

DISCUSSION

Extramedullary plasmacytomas manifest in the head and neck in more than 80% cases, especially in the nasal cavity or paranasal sinuses. In the literature, 32 plasmacytomas were identified from 15 studies with a primary site that was either the nasopharynx, clivus, sphenoid, or the petrous apex.[4,5,8] In a review of 50 orbital lesions, Mewis-Levin *et al.*,[9] noted that the orbit was more often involved with plasmacytoma associated with multiple myeloma than an isolated Solitary Plasmacytoma of Bone or Extramedullary Plasmacytomas. An exclusively orbital presentation of EMP is probably the first to be reported to the best of our knowledge. The features that lead to diagnosis are usually related to a functional disturbance created by the growth of the mass.

In this case, a gradual redness, swelling, and visual disturbances in the right eye were followed by progressive proptosis in the last 6 months. Establishing the diagnosis of plasmacytoma requires a biopsy. In this case, an incision biopsy was helpful to distinguish between several closely related pathologies, including plasma cell granulomas, plasmacytoid lymphomas, and large cell lymphomas (immunoblastic type). The histopathological slide was reviewed and immunohistochemistry was done, which confirmed the diagnosis of extramedullary plasmacytoma of orbit. The prognostic importance comes from a higher response rate to chemotherapy or radiotherapy, and a prolonged overall survival compared with those with the sole diagnosis of multiple myeloma.[3] Radiotherapy as the sole treatment option, as given in this case, is adequate in the absence of systemic involvement.

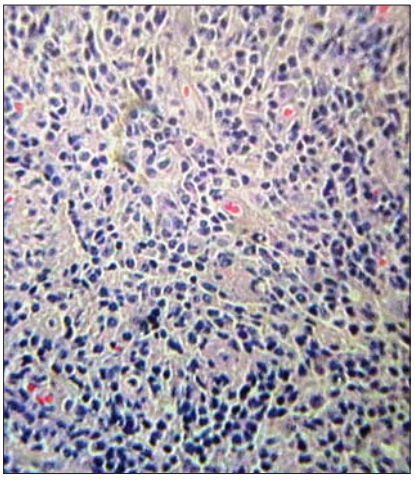

Figure 1: Histopathology from orbital soft tissue mass revealed plasma cell neoplasm

The biological behavior of plasmacytomas has been difficult to comment upon since most reports have been individual cases or small case series, *viz.* Bindal *et al.*, noted that petroclival infiltration was often associated with the development of multiple myeloma.[10] The likelihood of progression of a skull base Solitary Plasmacytoma of Bones (SPB) and EMP to multiple myeloma is unpredictable. Both can present with bone destruction but SPBs arising within the medullary cavity seems to have a higher risk as compared to EMPs, which, by definition, are extra-medullary, and mostly originates from a sub mucosal plane. In general, the likelihood of progression from isolated EMP to multiple myeloma is less than 30%, with a 10-year survival rate of 70%. For SPBs, the rate of progression is greater than 50%, with a 10-year survival rate of only 16%.[3,6] Till now, the available body of evidence is insufficient to comment whether these figures will hold true for solitary skull base plasmacytoma more so for the orbital ones.

Plasmacytomas have been classified as low grade, intermediate, or high grade using criteria originally intended for the grading of multiple myelomas**.** Chemotherapy was recommended after primary radiation therapy for the later two grades as these lesions had only a 17% local control rate with radiation alone. However, EMPs with low-grade histologies were controlled in 83% cases by external radiation alone. Our case being a low-grade EMP of orbital soft tissue and well-preserved visual acuity treatment with radiation alone was justified.

CONCLUSION

An orbital plasma cell neoplasm with no systemic involvement at present may be the first of its kind to be reported to the best of our knowledge. However, the risk of progression to multiple myeloma being not yet documented, should always be kept in mind, mandating a close follow-up of this patient.

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