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Extra Medullary Solitary Plasmacytoma of the Oropharynx: A Case Report

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Abstract

Background: Extramedullary plasmacytomas (EMP) are uncommon malignant neoplasms that can occur in any organ. They arise most frequently from the upper aerodigestive tract. It is necessary to exclude metastasis from multiple myeloma to diagnose a solitary extramedullary plasmacytoma. The pathophysiology of plasma cell neoplasms is related to the monoclonal plasma cell proliferation. Radiotherapy and surgery are the main treatments for EMP in the head and neck. The high risk of evolution toward myeloma encouraged new strategies for Solitary Plasmacytomas (SPs).

Materials and methods: Here we present a case study of a 60 year old male presented in our OPD with chief complaint of difficulty and pain while swallowing. On examination, we found a cystic swelling in left vallecula which was referred for excision. Histopathology was plasma cell neoplasm and immunohistochemistry consistent with the same. There were no lytic lesions and M band was absent. So, a diagnosis of extramedullary solitary plasmacytoma of oropharynx subsite vallecula was made. The post excision scans showed mild thickening of left vallecula and patient was started on bortezomib and lenalidomide based chemotherapy. The patient achieved complete response after 6 months of chemotherapy with minimal hematological and gastrointestinal toxicity.

Results: Most common sites of solitary extramedullary plasmacytoma of the head and neck include sinonasal area and nasopharynx. Here we reported a rare site of head and neck Solitary Extramedullary Plasmacytoma (SEP) i.e. vallecula (oropharynx). As the high risk of Myeloma is increasing in head and neck SEPs, new strategies have included bortezomib and lenalidomide in treatment of extramedullary plasmacytoma. Our patient achieved complete local response after excision and 6 months of chemotherapy.

Conclusion: As risks of Myeloma increase in solitary extramedullary plasmacytoma, Further similar studies will establish role of bortezomib and lenalidomide in extramedullary plasmacytoma. Currently surgery and radiotherapy form the mainstay of treatment.

Keywords: Cell proliferative disorder; Extramedullary plasmacytomas; Oropharynx; Lymph node

Introduction

Solitary plasmacytoma consists of a localized collection of malignant plasma cells without evidence of a systemic plasma cell proliferative disorder. These account for about 5% of all plasma cell neoplasms and may present with a single bone lesion (single bone plasmacytoma) or as a single extramedullary or extra-osseous lesion which is a rare presentation [1]. The pathophysiology of plasma cell neoplasms is related to the monoclonal plasma cell proliferation and the disease entity. The abnormal plasma cell clone can cause lytic lesions in the bone, inhibit normal hematopoiesis, and lead to deterioration of renal function through the deposition of abnormal immunoglobulin [2].

Extramedullary plasmacytomas are uncommon malignant neoplasms that can occur in any organ. They arise most frequently from the upper aerodigestive tract [3]. However, it is necessary to exclude metastasis from multiple myeloma to diagnose a solitary extramedullary plasmacytoma [4].

Extramedullary Plasmacytoma (EMP) often appears in the head and neck region. About 80% of EMPs occur in the submucosa of the upper aerodigestive tract. On both clinical presentation and pathologic examination, these tumors may be confused with some common tumors in the head and neck as they can also present with a swelling in the oral cavity, oropharynx, lymph node etc.

In a study, Clinical data of 10 consecutive patients with pathologically confirmed EMP in the head and neck were reviewed. The study concluded that the diagnosis of EMP mainly depends on clinical manifestation and pathologic results. Radiotherapy and surgery are the main treatments for EMP in the head and neck. Radiotherapy after surgical excision is standard treatment [5].

Here we are discussing a rare case of extramedullary solitary plasmacytoma of the vallecula and response to Velcade, Lenalidomide, Dexamethasone (VLD) based chemotherapy. The high risk of evolution toward myeloma encouraged new strategies for solitary plasmacytoma. Lenalidomide is a thalidomide derivative. It exerts direct

antitumor effect (apoptosis induction), immunomodulatory action, and antiangiogenic activity. Lenalidomide plus dexamethasone was shown superior to placebo plus dexamethasone in myeloma patients [6]. Bortezomib inhibits proteasome and thus prevents degradation of proapoptotic factors. Combination of bortezomib with melphalan-prednisolone was shown superior to melphalan-prednisolone alone in patients with diagnosed myeloma who were ineligible to high-dose therapy [7]. The use of bortezomib, with or without dexamethasone, could be a reliable, safe, and effective alternative for treating extramedullary plasmacytomas. In a study, sixteen months after diagnosis, the patient was in complete remission with no evidence of local relapse or evolution to multiple myeloma [8].

Case presentation

70 year male presented with difficulty in swallowing and pain in throat. On Indirect Laryngoscopy (IDL) examination, a cystic swelling was seen and palpated in the left vallecula. On RL (Rigid Laryngoscopy), a cystic lesion of around 3*2 cm was seen in left vallecula which was completely excised and sent for histopathology.

Histopathology report showed fibroconnective tissue densely infilterated sheets of mature and immature plasma cells, few binucleated cells were also noted. Impression was plasma cell dyscrasia. IHC (Immunohistochemistry) was done which showed Positivity for CD 138, MUM1, Kappa, CD79a and negative for CD 56, lambda, CD 20, PAX 5. The impression was plasma cell neoplasm (Kappa restricted). M spike was not seen in serum electrophoresis. Bone marrow studies showed plasma cell neoplasm with plasma cells less than 5%.

Low Dose whole body showed no bony lytic lesions. A diagnosis of solitary plasmacytoma of the oropharynx was made. Post excision IDL showed no residual but only pooled secretions in vallecula. CT showed minimal thickening at the left base of tongue area.

Treatment

Patient is currently on VLD (bortezomib, lenalidomide, and dexamethasone) regimen and is doing well with no reccurence post 6 months of excision (Figures 1-3).

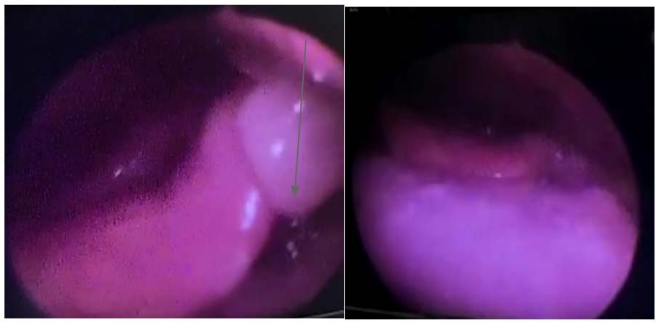

Figure 1: Showing rigid laryngoscopy finding on the left-Pre excision-on presentation, cystic nodule in left vallecula causing dysphagia and odynophagia. On the right-Post excision - post 1 month of excision-no swelling, only secretions at post excision site.

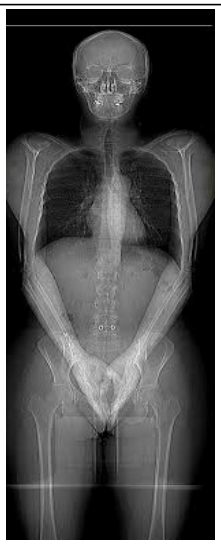

Figure 2: Showing low dose CT scan of the patient with no evidence of lytic lesions.

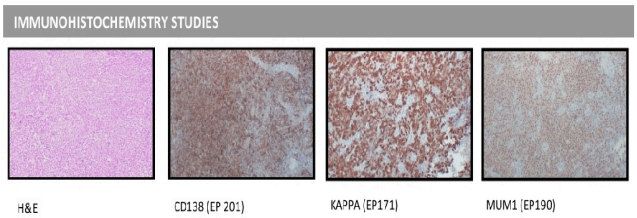

Figure 3: Showing immunohistochemistry slides H and E, EP201 showing CD138 positivity, EP171 showing Kappa positivity, EP 190 showing MUM 1 positivity.

Results and Follow Up

The patient has been on 6 months of chemotherapy VLD regimen (velcade/bortezomib, lenalidomide, dexamethasone) with regular follow up and with no fresh complaints.

The patient has not suffered from hematological toxicity or gastrointestinal toxicity. We also analyzed the clinical, biological, and radiological response to the treatment. Radiological responses were defined as complete response, partial response, or stable disease according to the Response Evaluation Criteria in Solid Tumors. It is a complete response until now.

Discussion

Solitary plasmacytoma consists of a localized collection of malignant plasma cells without evidence of a systemic plasma cell proliferative disorder. It accounts for about 5% of all plasma cell neoplasms and may present with a single bone lesion (single bone plasmacytoma) or as a single extramedullary or extra-osseous lesion[1]. Extramedullary Plasmacytoma (EMP) is a rare disease and is histopathologically characterized by infiltrates of plasma cells of diverse maturity and by their monoclonal immunoglobulin products [8].

The disease occurs almost exclusively in the head, neck, and upper respiratory tract. EMPs in the gastrointestinal organs are uncommon [9]. The next most frequent site of lesion occurrence is the stomach; however, this is also extremely rare, accounting for less than 5 % of all EMPs [10]. Although plasmacytoma is rare and few cases have been reported before, the adjuvant treatment lacks definitive guidelines. In such a situation we attempted with this unique combination therapy and have achieved satisfactory result.

The use of bortezomib, with or without dexamethasone, could be a reliable, safe, and effective alternative for treating extramedullary plasmacytomas [8].

Here we present a case study of a 60 year old male presented in our OPD with chief complaint of difficulty in swallowing and pain while swallowing. On examination, we found a cystic swelling in left vallecula which was referred for excision. On rigid laryngoscopy, a 3*2 cm cystic swelling was found in left vallecula. Histopathology was plasma cell neoplasm and immunohistochemistry consistent with the same. There were no lytic lesions and M band was absent. So, a diagnosis of extramedullary solitary plasmacytoma of oropharynx subsite vallecula was made. The post excision scans showed mild thickening of left vallecula and patient was started on bortezomib and lenalidomide based chemotherapy. The patient achieved complete response after 6 months of chemotherapy with minimal hematological and gastrointestinal toxicity.

Conclusion

Extramedullary Plasmacytoma (EMP) often appears in the head and neck region. About 80% of EMPs occur in the submucosa of the upper aerodigestive tract. Surgical excision followed by radiotherapy and chemotherapy. The role of bortezomib, lenalidomide, dexamethasone is established in solitary plasmacytoma. As risks of Myeloma increase in solitary extramedullary plasmacytoma, further similar studies will establish role of bortezomib and lenalidomide in extramedullary plasmacytoma. Currently surgery and radiotherapy form the mainstay of treatment.

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