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EXTRAMEDULLARY PLASMACYTOMA: A CASE REPORT

Radiotherapy

Dr. Rahul Mahawar* Junior Resident, Department of Radiation Oncology, RIMS, Imphal, Manipur, India. *Corresponding Author

Dr. L. Jaichand Singh Professor, Department of Radiation Oncology, RIMS, Imphal, Manipur, India.

ABSTRACT

Extramedullary Plasmacytoma is a rare, low-grade lymphoma which is mainly found in the head and neck region1. The diagnosis is usually made by exclusion. The paper reports a case of a 57-year-old male patient who presented with a single swelling in the lower part of anterior neck extending to upper part of thoracic region with a previous history of NHL. On palpation, a large mass (12 × 15 cm), hard, non-tender, fixed to the skin and underlying sternum was found in the neck and upper part of thoracic cavity. Trucut biopsy of the mass showed features suggesting EMP with IHC positive for CD138 and kappa. Chemotherapy and local radiation therapy were given to the patient and he is currently under follow-up.

KEYWORDS

Plasmacytoma, Lymphoma, Chemotherapy.

INTRODUCTION

Monoclonal extramedullary plasmacytoma (EMP or extraosseous plasmacytoma) is a rare, low-grade lymphoma which is mainly found in the head and neck region[1]. Its annual incidence is only 0.04 cases per 1,00,000 population[2]. It constitutes only 0.5% of all upper respiratory tract malignancies. It is more common in male patients. Male to female patients' ratio is 3:1 to 4:1[3]. It is most common in the head and neck region and mainly involves the nasopharynx, nasal cavity, paranasal sinuses and tonsils. Patient presents with complains of nasal obstruction, local pain, swelling or epistaxis. EMP is diagnosed on the basis of immunohistochemistry results and after the exclusion of systemic plasma cell proliferative disorder (multiple myeloma)[3]. A multimodality approach is used in the treatment of EMP.

Case Presentation

A 57-year-old male patient with history of hypertension and Type 2 diabetes mellitus came to the Department of Radiation Oncology, RIMS, Imphal with complain of swelling in the lower part of anterior neck extending to upper chest for last 1 year. There is no history of chest pain, cough, hemoptysis, fever, weight loss, night sweats or respiratory difficulty. The swelling was initially small but gradually increased in size over the course of 1 year to attain a size of roughly more than 10 × 10 cm. On palpation, a single large mass (12 × 15 cm), hard, non-tender, smooth margin, regular surface, fixed to the skin and underlying sternum was found in the lower part of anterior neck extending to upper part of thoracic cavity. No peripheral lymph nodes were clinically palpable.

The patient was diagnosed in May 2017 as a case of Low-grade B-NHL IVB (Extra nodal Marginal Node Lymphoma) - right lung mass with CD20 Positive, for which he received 6 cycles of chemotherapy with Bortezomib and Rituximab. Later, in April 2020 the patient developed a suprasternal swelling and core needle biopsy from the suprasternal soft tissue swelling showed features suggestive of Plasmacytoma with CD138 Positive and bone marrow biopsy showing Nodal Marginal Zone Lymphoma and CD20 Positive. He received 6 cycles of chemotherapy with Bortezomib and Rituximab from 4/7/20 to 10/12/20 for lymphoma and showed complete clinical response to treatment for lymphoma.

After the completion of lymphoma treatment, trucut biopsy of the neck mass was done which showed features suggestive of Extramedullary Plasmacytoma (with infiltration into adipose and skeletal muscles and mitotic activity 10-15/HPF) with CD20, CD56 & Lambda Negative and CD138 & Kappa Positive. Following this the patient came to the OPD of Department of Radiation Oncology, RIMS, Imphal on 29/1/2021.

Routine baseline investigations were done and were within normal range, Chest X Ray showed a single large mass in the lower part of neck extending to upper part of thoracic cavity. CT scan of neck and thorax showed a large soft tissue mass lesion (10.6 × 10.1 × 11.4 cm) on left cervical region extending downward up to left supraclavicular region and suprasternal area with compression of the left thyroid lobe and adjacent cervical soft tissue as seen in Figure . Bone marrow trephine biopsy showed plasma cells less than 2% with CD20 Negative.

The patient was planned for local radiation therapy and chemotherapy. The patient received external beam radiation therapy with Cobalt-60 teletherapy machine, to a total tumor dose of 5000cGy in 25#, five days a week for five days, in supine position by two parallel opposed fields (AP & PA) with spinal cord sparing after 40Gy. The patient was also started on chemotherapy with injection Bortezomib (D1,4,8 and 11) and tablet Lenalidomide (D1-28) for a total of 6 cycles (4 weekly cycle). The patient has completed his treatment and showed no evidence of EMP, and is now on regular follow up for the last six months.

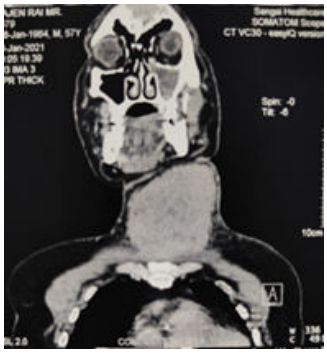

Fig A: CT neck and thorax (coronal view)

DISCUSSION

Solitary plasmacytomas are rare tumors of plasma cell origin (constituting only 4% of all plasma cell tumors). Immunophenotyping techniques are used to differentiate between EMP and benign polyclonal plasma cell proliferation. EMP is common between the age of 40 to 70 years[4]. EMP of the head and neck area, due to their clinical nature is considered a separate entity.

Diagnosis of solitary EMP is made after all the tests for disseminated disease are negative, which should include no signs of serum urine monoclonal protein, anemia, hypercalcemia or renal impairment. X-ray and/or MRI of the spine, pelvis, femurs and humerus should be normal. Bone marrow biopsy should also be within the normal range. The usual criteria for the diagnosis of solitary plasmacytomas (medullary or extramedullary), includes a biopsy-proven plasma cell tumor with one or at the most two solitary foci, Hb ≥ 13mg/dl, no Bence Jones protein in the urine, bone marrow taken at a distance from the primary site which is not involved by the tumor showing < 10% of plasma cells, and a normal serum electrophoresis at the time of diagnosis.

Metastasis to cervical lymph nodes is not common and varies with the site of the primary lesion. The exact relationship between EMP and multiple myeloma is not known, but approximately 20-30% of EMP cases will convert to multiple myeloma[3,5]. Due to low chance of local recurrence, treatment of choice for pedunculated EMP lesions is surgery, but for all other lesions it is radiation therapy alone or combined with other modalities[6,7]. A total tumor dose of 45-50Gy is indicated for effective local tumor control, but for patients who had extensive disease, a higher dose (50-60Gy) is used[8]. The risk of local recurrence is high, if a patient is treated with a tumor dose of less than 30Gy and only a minor risk for those treated with a dose of 40Gy or above.

CONCLUSION

Extramedullary plasmacytoma is a rare tumor of plasma cell origin whose diagnosis is very difficult and requires strict staging criteria, careful microscopic and immunohistochemistry studies for the correct diagnosis as it can be easily confused with other malignancies, like lymphoma. This study showed a case of 57-year-old man with EMP, who had shown complete response to radiation therapy and chemotherapy as seen in Figure B and C, and is currently under follow up.

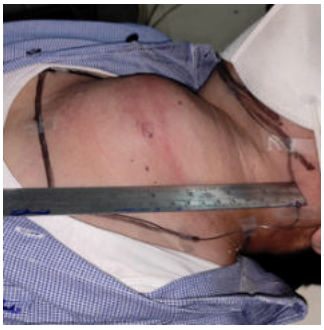

Fig B:Pre-treatment image of neck swelling

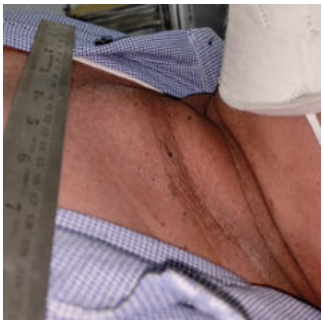

Fig C: Post-treatment image of neck swelling

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