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CASE REPORT

RADIOTHERAPY IN EXTRAMEDULLARY SOLITARY PLASMACYTOMA OF THE NASOPHARINX

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Abstract. Plasmacytomas are monoclonal proliferation of plasma cells. Extramedullary plasmacytoma (EMP) is a rare tumor (3-4% of all plasma cell tumors) that forms in soft tissues. The most common site for EMP is the head and neck (HN) region representing 0,4% of all HN malignant tumors. There are no set guidelines for the management of this disease due to it's rarity. For most of the cases radiotherapy is the standard treatment. We present a case of a 41 years old female diagnosed with extramedullary solitary plasmacytoma of the nasopharinx.
Keywords: extramedullary plasmacytoma, nasopharinx, radiotherapy

Introduction

Plasmacytomas are monoclonal proliferation of plasma cells. Most of the plasma cell tumors (multiple myeloma and solitary bone plasmacytoma) arises in the myeloid tissue of the bone. Extramedullary plasmacytoma (EMP) is a rare tumor (3-4% of all plasma cell tumors) that forms in soft tissues, characterised be the presence of focus of monoclonal plasma cells in the soft tissue in the absence of systemic disease. EMP was first described by Schridde et al in 1905. [1,3,4]

The most common site for EMP is the head and neck (HN) region representing 0,4% of all HN malignant tumors, more frequently in paranasal sinuses, nasal fossa and nasopharinx. Symptoms are related to local tumor effects: epistaxis, nasal obstruction, pain, anosmia, nasal congestion. For the diagnosis of EMP it is necessary to exclude multiple myeloma, by performing serum protein electrophoresis, urinalysis for Bence-Jones protein (rarely seen in EMP), skeletal radiological assessment and bone marrow biopsy (less then 5% plasma cells in the bone marow in EMP). Differential diagnosis should exclude benign tumor, carcinoma, lymphoma. The clinical behavior can be transformation to multiple myeloma, local recurrence, distant metastasis. [2,5,6,7]

There are no set guidelines for the management of this disease due to it's rarity. Surgical management can achieve local control, but it is limited to the cases when radical excision is possible. There are no data that chemotherapy as primary treatment improve survival or reduce relapses in EMP. Plasmacytomas are radiation sensitive therefore the main treatment in EMP is radiotherapy. Doses varies in the range of 40 to 60 Gy over 4 – 6 weeks with a threshold of 40 Gy for local control. The policy of nodal irradiation remains controversial but there are data arguing in favour of treating only the primary tumor and involved nodes. [9,10,11]

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Patient, Methods and Results

We present a case of a 41 years old female with personal and family history N/A. She was evaluated in a otorhinolaryngology service for progressive nasal obstruction, epistaxis and discharge. After initially surgery for adenoids, the symptoms where persistent, further investigations showed a tumor in the nasopharinx. A biopsy was performed and the morphology & immunophenotype confirmed the diagnosis of a plasmacytoma. The serum protein electrophoresis showed increased value of IgA , the bone marrow biopsy confirmed less then 5% plasma cells and the skeletal survey didn't fiind any abnormalities. Head and neck MRI noticed the thickening of the nasopharinx mucosa, with no enlarged lymph nodes.

The final diagnosis was extramedullary solitary plasmacytoma of the nasopharinx. Additional workup include thoracic CT, posterior rhinoscopy, abdominal ultrasound, laboratory tests.

After submitting the case to the tumor board (head and neck surgeon, haematologists, radiation oncologist) the recommendation was radiotherapy.

We performed 3D conformal radiotherapy with CT planning. For positioning and immobilization the patient was supine with neck extended, with the use of an aquaplast mask to immobilize head and shoulders. CT scan was performed with i.v. contrast, 3 mm thick slices, from vertex to aortic arch. For treatment volume delineation we choose the CTV including the whole nasopharinx, according to the data that plasmacytoma frequently disseminates by submucosal way. There was no GTV delineated (in this case difficult to delineate a tumor with only thickening of the nasopharinx mucosa on MRI and CT). We choose not to include the nodal areas. (fig.1)

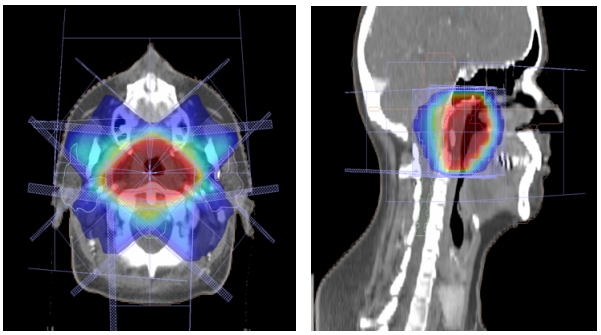

Figure 1. CT axial and sagittal slice showing CTV (red), PTV (pink) and dose distribution (color wash)

The organs at risk was also delineated: optical structures, parotids, brain stem, spine, pituitary, temporomandibular joints, cochleas. Verification and approval of the plan was made after checking the dose distribution and the dose volume histogram. (fig2)

For treatment delivery we used a linear accelerator,

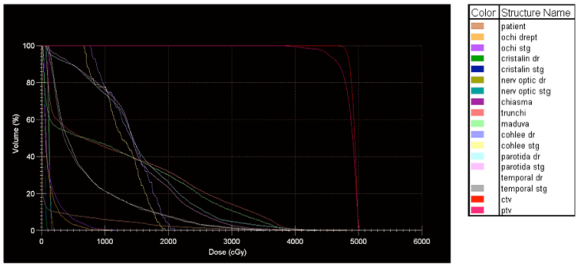

Figure 2. Dose volume histogram

with multiple 6 MV photons beams. Verification was performed with portal imaging system, with images taken on days 1-3 and then weekly. The dose/fractionation schedule was 2 Gy/ fraction, 1 fraction/ day, 5 days/ week, to a total dose of 50 Gy in 25 fractions over 5 weeks.

During treatment the patient experienced acute side effects: grade 1 dermatitis and dysphagia.

Follow up after treatment for 2 years (clinical examination, MRI, rhinoscopy) showed no evidence of disease.

Discussion

Extramedullary solitary plasmacytoma of the nasopharinx is usually treated with radiotherapy. The role of surgery remains limited due to the impossibility to perform radical excision. Chemotherapy didn't prove to reduce relapses or improve the survival.[12,13,14]

There are limited data regarding radiation treatment in EMP of the head and neck region, with no prospective studies and only few retrospective studies. Doses in the range of 40 to 60 Gy usually achieve good local control. There is a controversy regarding the role of elective nodal irradiation.[12]

Long term folow up is recommendable due to data showing progression to multiple myeloma even 15 years after completion of radiotherapy.[12,13,14]

In our case we choose 3D conformal radiotherapy to a total dose of 50 Gy in 25 fractions over 5 weeks, treating only the nasopharinx with no elective nodal irradiation. The treatment was well tolerated. The patient is free of disease after 2 years.

Conclusion

Due to its rarity, extramedullary solitary plasmacytoma of the nasopharinx poses a challenge for physicians, regarding diagnosis and treatment. There are no set guidelines for the management of the ESCCs and prospective trials are warranted.

References

1. **Schridde H.** Weitere Untersuchungen uber die Kornelunger der Plasmazellen. *Centralbl Allg Pathol Anatol.* **1905; 16: 433-5.**

2. **Wiltshaw E.** The natural history of extramedullary plasmocitoma and its relation to solitary mieloma of bone and myelomatosis. *Medicine.* **1976; 55: 217-38.**

3. **Kapadia SB, Desai U, Cheng VS.** Extramedullary plasmacytoma of the head and neck: a clinicopathologic study of 20 cases. *Medicine* **1982; 61: 317–29.**

4. **Susnerwala SS, Shanks JH, Banerjee SS, Scarfee JH, Farrington WT, Slevin NJ**, Extramedullary plasmacytoma of the head and neck region: clinicopathologic correlation in 25 cases. *Br J of Cancer*; **1997; 75: 921-927.**

5. **Nofsinger YC, Mirza N, Rowan PT, Lanza D, Weinstein GS**, Head & neck manifestations of plasma cell neoplasms. *Laryngoscope*; **1997: 107: 741-746.**

6. **Bachar G et al.** Solitary extramedullary plasmacytoma of the head and neck--long-term outcome analysis of 68 cases. *Head Neck.* **2008 Aug; 30(8): 1012- 9**.

7. **Kilciksiz et al.** Clinical and prognostic features of plasmacytomas: a multicenter study of Turkish Oncology Group-Sarcoma Working Party. *Am J Hematol.* **2008 Sep; 83(9): 702-7.**

8. **Mendenhall CM, Thar TL, Million RR.** Solitary plasmacytoma of bone and soft tissue. *Int J Radiat Oncol Biol Phys.* **1980; 6: 1497–501.**

9. **Alexiou C, Kau R, Dietzfelbinger H, et al**, Extramedullary Plasmacytoma tumor occurrence and therapeutic concepts. *Cancer*; **1999; 85: 2305-2314.**

10. **Soutar R, Lucraft H, Jackson G, et al**, Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. *British J of Haematology* **2004; 124: 717-726.**

11. **Kumar S.** Solitary plasmacytoma: is radiation therapy sufficient? *Am J Hematol.* **2008 Sep; 83(9): 695-6.**

12. **V J Michalaki.** Definitive radiotherapy for extramedullary plasmacytomas of the head and neck,

The British Journal of Radiology, **76 (2003), 738–741.**

13. **Soesan M, Paccagnella A, Chiarion-Sileni V, Salvagno L, Fornasiero A, Sotti G, et al.** Extramedullary plasmacytoma: clinical behaviour and response to treatment. *Ann Oncol* **1992; 3: 51–7.**

14. **Alexiou C, Kau RJ, Dietzfelbinger H, Kremer M, Spiess JC, Schratzenstaller B, et al.** Extramedullary plasmacytoma: tumor occurrence and therapeutic concepts. *Cancer* **1999; 85: 2305–14.**

15. **Jyothirmayi R, Gangadharan VP, Nair MK, Rajan B.** Radiotherapy in the treatment of solitary plasmacytoma. *Br J Radiol* **1997; 70: 511–6.**