

Nasal Plasmacytomas: Report of Two Cases and Revision of the Literature

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Abstract

Solitary plasmacytomas (SP) represent less than 1% of head and neck (HN) tumors. The most frequent initial presentation in HN of SP is at the naso-sinusal and nasopharynx levels (75%). The article present two cases of naso-sinusal SP treated at our Center recently with endoscopic surgery followed by radiation therapy, focusing in the combined modality treatment for this disease.

Keywords: Nasal Plasmacytoma; Endoscopic surgery; Radiation therapy

Introduction

Solitary plasmacytoma is a rare disorder, similarly as myeloma. The patient diagnosis with solitary plasmacytoma don't have myeloma cells within the bone marrow or throughout the body instead they the tumor is composed of plasma cell. The reason for solitary bone plasmacytoma isn't found yet because it's presentation in mucosa of aerodigestive tract and etiology of extramedullary plasmacytoma is said to chronic stimulation of virus infection.

Case Report

Case 1

An 83-year-old farmer, with a history of COPD, ischemic heart disease and claudication of the lower extremities, who visited

the otolaryngologist in January 2018, reporting symptoms of nasal respiratory failure and hyposmia of months of evolution. Nasopharyngoscopy revealed a pedunculated mass in the right nostril (RN) with a neoplastic appearance that occupied the entire fossa and seemed to depend on the inferior or middle turbinated bone with occupation of the meatus. Magnetic Resonance Imaging (MRI) (**Figure 1**) showed a mass in the RN, centered in the middle meatus, with expansion and bone remodeling of the nasal septum and the lamina papyracea of the right ethmoid, suggestive of neoplasia. The biopsy revealed a diffuse, high-grade monomorphic proliferation, poorly differentiated, with a high mitotic index, expressing positivity for vimentin and plasma markers (CD38 and CD138), compatible with the diagnosis of plasmacytoma of the plasmablastic phenotype (sphenoid sinus and right nostril). On January 2018 he underwent surgery through nasosinusal endoscopic surgery, showing a tumor that occupied the entire RN, dependent on the middle turbinated bone with extension to the middle meatus and upper edge of the lower turbinated bone, reaching the anterior ethmoid. Surgery consisted of macroscopic tumor resection of the tumor. The pathological study confirmed the diagnosis of plasmacytoma. The patient was studied by the Service of Hematology, where they requested a complete blood analysis, a bone marrow aspirate and a PET scan, ruling out the existence of multiple myeloma (MM). Treatment was completed with Intensity Modulated Radiotherapy (IMRT) on the tumor bed with a 5 mm margin, administering a total of 44 Grays in 22 sessions through 6 fields (**Figure 2**). Subsequently, the patient has continued check-ups without signs of recurrence, the last 34 months after diagnosis.

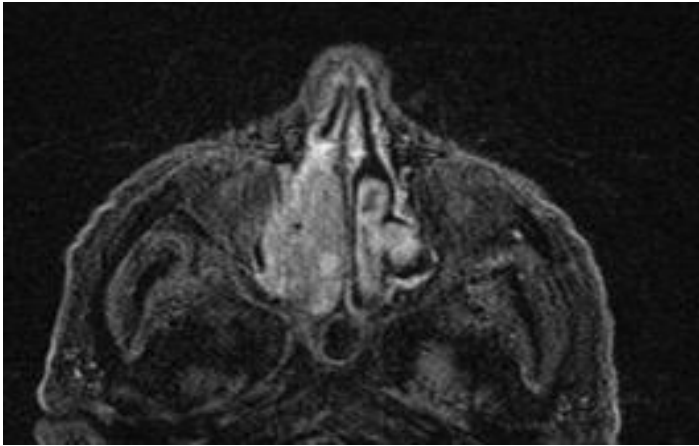


Figure 1 NMR of patient number 1, showing a mass in the right nostril centered on the middle meatus suggestive of neoplasia in the right naso-ethmoid region with possible involvement of the ipsilateral maxillary sinus.

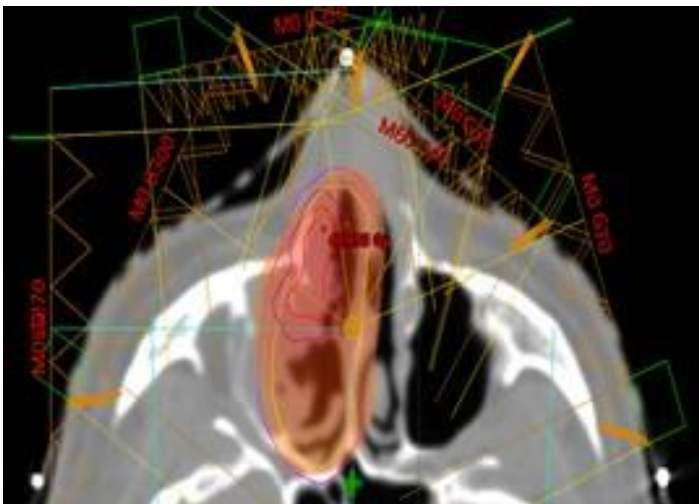


Figure 2 Image of the RT plan of patient number 1 (6 IMRT fields). 95% isodose in color.

Case 2

A 64-year-old patient, ex-smoker and moderate drinker, with a personal history of arterial hypertension, prostate hypertrophy, Peyronie's disease, ascending aortic aneurysm. He went to the Otolaryngology Service in April 2017 for slight bleeding in RN after daily removal of a crusty lesion in RN. He did not report pain or other discomfort. Nasopharyngoscopy revealed a small mass in the inferior turbinated bone covered with crust. Computed Tomography (CT) (**Figure 3**) showed a soft tissue lesion that seemed to depend on the inferior turbinated bone and nasal septum, with greater contact with the lateral wall. On July 2017, he underwent nasosinusal endoscopic surgery, showing a lesion with a neoplastic appearance that depended on the body of the inferior turbinated bone and the lateral wall of the RN, performing a macroscopic resection of the lesion and the inferior turbinated bone. The histological study showed fragments of respiratory mucosa, in the chorion of which a monomorphic proliferation of

cells was identified, with a characteristic morphology of a plasma cell, densely cellular. This cell proliferation strongly expressed plasma cell markers, such as CD138, being negative for CD20 and CD56. There was a monotypic expression for the Kappa light chain, all consistent with a plasmacytoma. The patient was subsequently studied by the Hematology service where, after the pertinent explorations including PET, the presence of a MM was ruled out. Given the diagnosis of a stage IA nasal plasmacytoma with incomplete surgery (R1), it was decided to complete the treatment with Intensity Modulated Radiotherapy (IMRT) on the tumor bed with a margin of 5 mm, administering a total of 40 Grays in 20 sessions through of 4 fields (**Figure 4**). After treatment, the patient continued check-ups without signs of recurrence, 44 months after diagnosis.

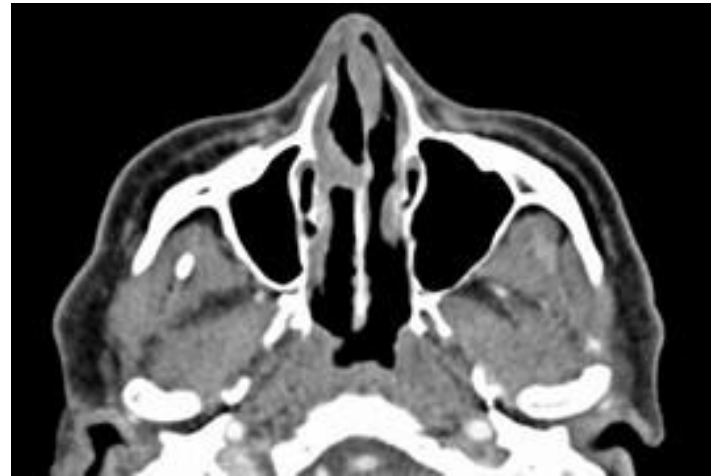


Figure 3 CT of patient number 2 showing soft tissue lesion that contacts the inferior turbinated bone and nasal septum, with greater contact with the lateral wall.

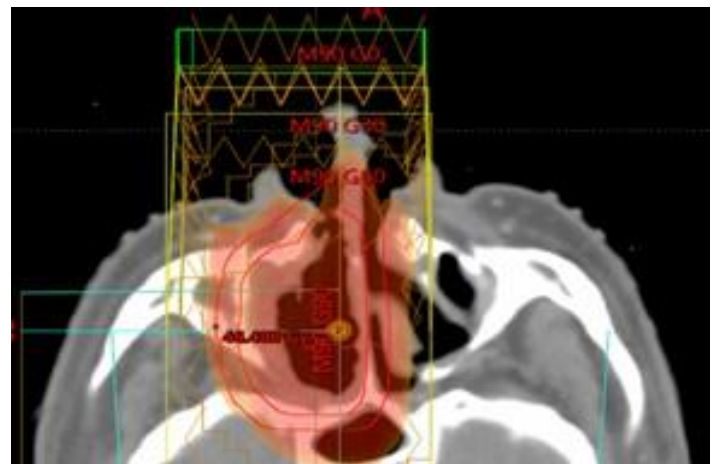


Figure 4 Patient number 2 (4 IMRT fields).

Discussion

Solitary plasmacytomas (SP) represent less than 1% of head and neck (HN) tumors. They mainly affect males in a 4:1 ratio [1]. The most frequent initial presentation in HN of SP is at the nasosinusal

and nasopharynx levels (75%), followed by the oropharynx (12%). The most common symptom in those with sinonasal location is upper respiratory tract obstruction, followed by epistaxis (since they are highly vascularized tumors) [2]. The presence of positive neck nodes is variable (0-25%). The diagnosis of SP is histological by biopsy of the tumor [3]. Plasmacytomas are neoplasms of immunoglobulin-secreting plasma cells, and in 80% of cases they are well defined. The histological diagnosis is established by the presence of aggregates of plasma cells with frequent atypia such as the presence of irregular nucleoli, inversion in the nucleus/cytoplasm ratio, mitosis and infiltration of adjacent tissue. The monoclonal nature of plasma cells is confirmed by immunocytochemistry (immunoperoxidase) [3]. Inflammatory findings, including granulomas and amyloid deposits, are common. The pathological examination allows us to reach the diagnosis of plasma cell tumor, but there are no pathognomonic findings that allow us to differentiate between SP and MM [3] indeed, the diagnosis of MM is based on three clinical findings:

- a) Histological evidence of plasmacytoma or plasmacytosis in bone marrow.
- b) Clinical evidence of disease such as bone pain, anemia, or kidney failure.
- c) Monoclonal gammopathy in serum or urine or osteolytic lesions. Regarding treatment, PS is highly radiosensitive tumors [4-7], the results being similar with radiotherapy or surgery, both producing excellent local and regional control.

In nasosinusal tumors, a complete excision is not usually performed due to their proximity to vital structures, which is why most will require adjuvant radiotherapy [8]. The irradiation dose is not well defined due to the small number of patients and the lack of prospective studies, although in general doses around 40-50 Grays in 20-25 fractions are recommended [9]. The adjuvant treatment with CT (melphalan and prednisone) to prevent progression to MM is controversial. 5-year disease-free survival ranges between 50% and 90% in most authors [10]. Transformation to MM occurs more frequently in nasosinusal plasmacytomas (up to 40% depending on the series).

Conclusion

The strategy of endoscopic surgery and postoperative radiotherapy constitutes a highly effective treatment with excellent local control in solitary naso-sinusal plasmacytomas, the transformation to MM being the main cause of mortality.

Conflicts of interest

The authors declare no conflicts of interest regarding the publication of this paper.

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