Plasmacytoma an Infrequent and Particular Tumor of Nasal Cavity: A New Case Report

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Abstract

We report a new case of extra-medullary solitary plasmacytoma of the nasal cavity in order to emphasize this rare tumor with a predilection for sinuses and nasal cavity levels and to expose its diagnostic and therapeutic features. It was a 60-years-old woman who consulted for a left-sided nasal cavity mass associated with an epistaxis. The clinical examination revealed a swelling of the left nasal region.

CT-scan showed a mass completely filling the maxillary sinus, ethmoidal cells, frontal sinus and sphenoidal sinus. The cervico-facial MRI showed a soft tissue mass completely filling the left maxillary sinus enlarging the ostium, the osteomatous intersection and the left nasal cavity, associated with a frontal and ethmoidal sinusal reaction of the left anterior sinuses.

Histopathologically, it was a monoclonal plasma cells proliferation. Skeletal imaging confirmed the absence of bone localization. Results of serum electrophoresis and urine tests were negative for myeloma component or Bence-Jones protein. The diagnosis of extramedullary plasmacytoma of the nasal cavity was therefore retained. Extramedullary plasmacytoma of the nasal cavity is rare and should be considered in the differential diagnosis of nasal cavity masses especially in young age group.

Keywords: Nasal; Plasmacytoma; Solitary; Extra-medullary

Introduction

Extra Medullary Solitary Plasmacytoma (EMSP) is a rare tumor which represents less than 1% of head and neck cancers and less than 0.4% of Upper Respiratory Tract (URT) cancers. These tumors have a predilection for sinuses and nasal cavities [1]. EMSP of nasal cavity is a monoclonal proliferation limited to mucosa out of medullar impairment. Certain diagnosis is made on histological examination. For localized forms, radiation therapy is usually sufficient and considered as a reference treatment. In some cases it may be associated to surgery.

Case Report

We report the case of a 60-years-old woman with medical history of arterial hypertension and asthma. She consulted for a left nasal cavity mass with epistaxis. Clinical examination revealed a left nasal cavity tumor preventing airflow. Furthermore physical examination was normal particularly there were no cervical adenopathies. Anterior rhinoscopy showed a polypoid reddish proliferation filling out left nasal cavity. Cervico-facial CT-scan showed a soft-tissue mass that filled the left-sided meatus, with total opacity of the left maxillary, ethmoid, frontal and sphenoidal sinuses.

For better identification of the tumor a cervico-facial MRI was recommended. It objectified a blockage of the middle left-sided meatus by a soft-tissue proliferation with reactive obstruction of ipsilateral frontal and ethmoid sinuses. Multiple reactional polyps were seen in the right maxillary sinus. Left maxillary and ethmoid sinuses biopsies were made. Microscopic examination on hematoxylin-eosine staining showed a malignant proliferation made of broad sheets of plasma cells with a scanty connective tissue stroma. Cells had round to oval eccentric nuclei with cart-wheel chromatin and basophilic stained abundant cytoplasms (Figures 1-3).

Immunostaining showed positivity of tumor cells for CD79a and CD138 and Lambda chain monoclonality (Figure 4 and 5). A skeletal survey showed no medullar impairment. Serum protein electrophoresis showed normal levels of immunoglobulins and Bence Jones protein urine test was...
negative with normal renal function. The Diagnosis of EMSP was retained.

Our patient received a total dose of radiation therapy of 44 Gray for a fraction size of 2 Gray per session. She had a 30 months recovery. The tumor relapsed in February 2015. It was irradiated with positive evolution retrospectively.

Discussion

Plasmacytomas are a disparate group of affections. Myeloma is the disseminated form, however solitary plasmacytoma is the located form and which may be medullary or extra medullary. EMSP represent 10% of solitary plasmacytoma. They are mostly located in upper respiratory tract submucosa and account for 0.4% of head and neck malignant tumors [2].

In 25% of cases respiratory tract ESMP may extend to local lymph nodes and to other organs in 10% of Cases [3]. EMSP is a mucosal monoclonal plasma cell proliferation apart from medullary impairment and have better prognosis than disseminated forms [4].

These tumors originate from plasma cells of Mucosa Associated Lymphoid Tissue (MALT).

Etiopathogenesis of EMSP has been a long-standing issue. Chronic stimulation by irritating agents or viral infections generating antigenic stimulation was accused. Genetic factors and exposure to radiations were mentioned in literature as risk factors but none association between EMSP and these factors was practically established [5]. EMSP has a marked male predilection with a sex ratio of 4/1. It affects adults between 50 years and 60 years old.

Clinical presentation is not really characteristic. Epistaxis, nasal obstruction or congestion, serous or bloody nasal discharge and/or facial palsy may reveal the tumor. Pain is generally absent [3]. Physical examination reveals generally a pediculate or even sessile polypoid reddish, buff-colored or grey proliferation. Adenopathies must be searched for carefully. Imaging determines tumor size and local extension.

It’s compulsory to rule out tumor extension or multiple tumors to retain the solitary nature of the tumor [2]. Further investigation (Lab test and imaging) is a must [2]. It helps distinguishing solitary forms from systemic ones and medullary from extra medullary impairment [2].

Certain diagnosis is retained after histological examination. The tumor is made of broad sheets of plasma cells with a scanty connective tissue stroma. Cells had round to oval eccentric nuclei with cart-wheel chromatin and basophilic stained abundant cytoplasm. Giant cells may be seen occasionally [3]. Tumor cells have a B immunophenotype expressing CD79a and CD138 with Lambda or Kappa chain monoclonality. Once the diagnosis of EMSP is retained on a histological examination of a biopsy multiple myeloma should be ruled out by a negative skeletal imaging, absence of monoclonal gammapathies on protein electrophoresis and absence of Bence-
Jones proteins in urine [1].

In front of such lymphoid proliferation, the main differential diagnosis is Hodgkin lymphoma, in particular lymphoplasmacytic lymphoma. Immunohistochemistry is of a great help in such cases and allow straightening the diagnosis [2].

EMSP are radiosensitive [6-8]. Radiation therapy is the reference treatment. Nevertheless it remains a contentious issue concerning dose and target volumes. Local control with improved survival is evaluated between 80% and 100% with moderate doses of radiation therapy [9,10]. Surgery is commonly mutilating and exceptionally indicated since these tumors have excellent radio sensitivity [11]. However it may be indicated in a late stage in case of recurrence or progression after radiation therapy [12]. No evidence has been yet established about the efficiency of adjuvant chemotherapy in treating EMSP. The latter is suggested when radiation therapy is not available [3].

These tumors still have a better prognosis than disseminated forms. Still actually there are no retrospective studies providing data about overall survival and recurrence rates. However, Cases of local recurrence or dissemination many years after treatment have been reported and which justify a close and long-term follow up [3].

Conclusion

EMSP are infrequent tumors affecting preferably upper respiratory tract mucosa. The diagnosis of EMSP can be retained only once an eventual myeloma is ruled out even though its occurrence is rare in extramedullary forms compared to medullary ones, which justify the whole initial assessment and dictate a regular and close follow-up. Treatment is based on radiation therapy.

References