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REPORT OF EXTRAMEDULLARY PLASMACYTOMA OF THE NASAL CAVITY, A RARE ENTITY.

ENT

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ABSTRACT

Background: Extramedullary plasmacytoma is a rare neoplasm characterized by the monoclonal proliferation of plasma cells. The nasal cavities and nasal septum are the most common sites of occurrence. This entity typically presents in the 5th to 6th decade of life. **Materials And Methods:** We present the case of a 74-year-old male with a 2-month history of progressive right-sided nasal obstruction and self-limiting epistaxis. He had a 40-year history of smoking and social alcohol use. Ear, nose, and throat (ENT) examination and anterior rhinoscopy revealed a smooth, pale-reddish mass in the right nasal cavity, adhered to the lateral wall and insensitive to probing. **Results:** Computed tomography (CT) of the sinuses revealed a soft tissue mass occupying the right nasal cavity extending to the choanae. Biopsy with hematoxylin and eosin staining showed diffuse plasma cells separated by vascular septa. Immunohistochemical analysis demonstrated a slight increase in kappa light chain expression. Hematologic evaluation ruled out multiple myeloma, confirming the diagnosis of extramedullary plasmacytoma. The tumor was completely excised transnasally via endoscopy, followed by radiotherapy. **Conclusion:** Extramedullary plasmacytoma is a rare, potentially aggressive tumor requiring a multidisciplinary approach for diagnosis and management. Long-term follow-up is essential to monitor for recurrence or progression.

KEYWORDS

Nasal cavity, plasmacytoma, surgical excision, radiotherapy.

INTRODUCTION

The term extramedullary tumour was first described in 1905 by Schridde. The estimated global incidence of the disease is 1 case per 500,000 people[1,2]. Extramedullary plasmacytoma corresponds to less than 10% of all plasmocytic tumors[1,2], representing less than 1% of all head and neck tumors[4] and less than 0.5% of tumours of the aerodigestive tract[3].

It is important for the otolaryngologist to acquire knowledge of this disease since 80 to 90% of the extramedullary plasmacytoma cases are found in the head and neck[4]. This disease occurs in people above 40 years of age (over 95% of cases)[4], typically between the sixth and seventh decades of life, affects 3 to 4 times more men than women, especially in Caucasians, and exhibits a slowly evolving nature.

Extramedullary plasmacytoma is a plasma cell tumour that forms in soft tissues such as the lymph nodes, skin, and mucosa; thus, by definition, this tumour has no origin in the myeloid tissue of the bone.

The tumour mass is not histologically distinguishable from myeloma. The tumour has a firm consistency, which can be sessile or pedunculated, the colour ranges from greyred to grey, and easy bleeding is often observed under manipulation.

When these tumours occupy the nasal cavities, a differential diagnosis should be conducted in order to rule out other bleeding tumours, especially squamous cell carcinomas. When determining the presence of a clone of plasma cells in the biopsy, an effective analysis must be performed to confirm the existing plasma cell disorder

MATERIALS AND METHODS

A 74-year-old male came to the Otorhinolaryngology department of a Tertiary Care Hospital with progressive right side nasal obstruction that had lasted for the last 2 months and was accompanied by self-limited epistaxis.

His background indicated a history of smoking (40 years packet) and social drinking. Upon Ear, Nose, and Throat (ENT) examination, a smooth reddish mass was observed on the right nasal cavity. Fig 1.

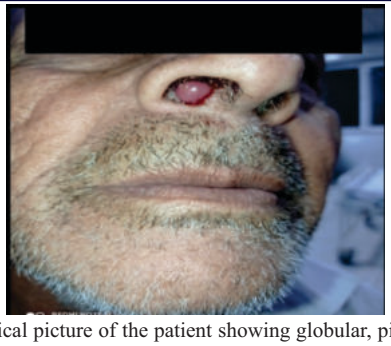

Fig 1- Clinical picture of the patient showing globular, pinkish mass protruding in right vestibule and occupying the right nasal cavity.

Anterior Rhinoscopy revealed a pale-reddish, smooth mass that occupied the right nasal cavity and apparently adhered to the side wall and insensitive to probing. Left nasal cavity is normal and patent with mild deviation of septum. Computed tomography (CT) of the sinuses revealed soft tissue attenuation occupying the right nasal cavity to the choanae. Fig 2.

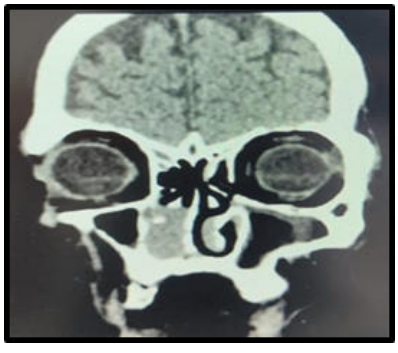

Fig 2- CT Paranasal sinuses (Plain + Contrast)- Coronal cut showing heterogeneously enhancing soft tissue mass occupying right nasal cavity extending into right middle turbinate and inferior turbinate

Incisional biopsy of a lesion of the right nasal cavity was performed under local anaesthesia. Haematoxylin and Eosin staining showed many plasma cells arranged in diffuse pattern separated by vascular septate. Fig 3.

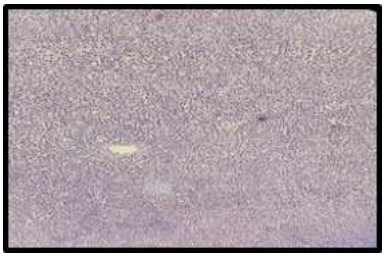

Fig 3- Haematoxylin and Eosin staining of plasmacytoma reveals mature and immature cells with moderate to abundant cytoplasm characterises eccentric nuclei

Immunohistochemical assays shown a slight rise in kappa chain. The patient underwent a thorough hematologic evaluation, including bone marrow biopsy, bone imaging, protein electrophoresis, and Bence-Jones proteinuria. This evaluation excluded multiple myeloma and demonstrated that his condition was extramedullary plasmacytoma.

Then complete surgical excision of tumour trans nasally with endoscope was done followed by radiotherapy. Patient is advised for follow-up every 6 months.

DISCUSSION

Plasma cells are mature immunocompetent cells derived from B lymphocytes, and they provide specific noncellular immunity within the immune system. They produce specific antibodies against antigens in different tissues. Large populations are seen in the mucosa of the nasal and paranasal sinuses. Plasmacytomas arises from a neoplastic proliferation of these antibody—producing plasma cells. [1] The three types of plasmacytomas – multiple myelomas, solitary plasmacytomas and extramedullary plasmacytomas are further classified as either localized (stage I), localize including local lymph nodes (stage II), or generalized (stage III) according to the clinical manifestations.[2]

A systemic method of staging of the disease – including measurement of the complete blood count, renal and liver functions, serum and urinary protein electrophoresis, and serum immunoglobulin levels; a skeletal survey and bone marrow examination; and computerized tomography of the tumour region – must be performed in order to exclude systemic involvement before the diagnosis can be made.[1]

CT scan of nose and para nasal sinuses (PNS) is done to evaluate spread of the disease.

Immunohistochemical staining will demonstrate the monoclonal nature of the plasma cells and confirm the neoplastic nature of the lesion. The search for light chains allows the determination of whether the process is monoclonal, i.e., KAPPA or LAMBDA chain. In addition, immunohistochemistry study is used to differentiate extramedullary plasmacytoma from benign reactive plasmacytosis as well as other malignant disorders, such as undifferentiated carcinoma, melanoma and esthesioneuroblastoma.[3] It also differentiates the inflammatory processes, which are rich in the plasma cells in this region.

A negative bone marrow biopsy, the absence of bone lysis, and normal electrophoresis of blood immunoglobulins allows the possibility of a multiple myeloma to be excluded.

Thus, a diagnosis can be made by observing the following features [2,4]:

1. Histological evidence of nasal mass
2. Bone marrow aspiration showing less than 10% atypical plasma cells
3. Absence of clinical and radiological evidence of skeletal lesions
4. Unchanged levels of serum proteins or electrophoresis of serum/urinary proteins
5. Absence of anemia (80–85% patients with multiple myeloma have normochromic normocytic anemia)

5-year survival rates for extramedullary plasmacytoma are between 30% and 80%. Most deaths occur from development of disseminated multiple myeloma. The rate of conversion of extramedullary plasmacytoma to multiple myeloma has been shown to be from 10 to 36% in various series. Progression to multiple myeloma usually occurs within 2 years of diagnosis, but has occurred up to 15 years later indicating the need for long-term follow-up. This biphasic nature of progression to multiple myeloma suggests that patients with early relapse probably had undetected myeloma present at the time of diagnosis. The more slowly progressing group may represent patients who developed new primaries in dysplastic marrow. This interpretation has important implications for more intensive screening to be carried out to exclude a diagnosis of multiple myeloma. [5]

Regarding the natural history of the disease, Batsakis [6] defined 5 possible stages that extramedullary plasmacytoma can present:

1. Localized disease; solitary; controlled by surgery, radiotherapy, or both; without recurrence; or disseminated.
2. Disease with local recurrence controlled by additional therapy
3. Aggressive disease, persistent or recurrent; death by uncontrollable local extensions.
4. Local disease with regional lymph node "metastasis" without evidence of distant spread.
5. Local disease, recurrent or followed by dissemination and development of another neoplasm of plasma cells and/or multiple myeloma.

Wiltshaw stated in his study that 40% of extramedullary plasmacytomas spread beyond the primary site and/or undergo lymphatic drainage. [7] Of these, 62% of patients had deposits in soft tissues and visceral organs, and 81% developed bone lesions. An extramedullary plasmacytoma at this stage is called Disseminated Extramedullary Plasmacytoma, and its prognosis is undoubtedly better than that of multiple myeloma. However, the possibility for conversion of extramedullary plasmacytoma to multiple myeloma exists. Its incidence rate varies from 15 to 20% and its prognosis is poor.[5]

Regarding the treatment of extramedullary plasmacytoma in the head and neck, the choice of radiotherapy or surgery alone is controversial in the published data. Some authors advocate the use of radiotherapy as a sole treatment due to the good response in most cases. Chemotherapy is indicated only in cases of disseminated disease. [8, 9] In 1993, Wax recommended surgery in cases of localized lesions that could be removed with minimal morbidity. [10]

Regarding nodal dissemination (which occurs rarely) and local relapse, controversy persists because although metastases respond with the same sensitivity to radiotherapy, isolated surgery may also be indicated. In 1988, Abemayor recommended the exclusive use of radiotherapy and suggested reserving surgery only to remove residual disease. [11] Although it is known that extramedullary plasmacytoma is a radiosensitive tumour, no consensus exists in the literature regarding the optimal dose for therapy.

Regarding the results of the possible treatments, in 1976, Wiltshaw obtained a local recurrence rate of 21% for radiotherapy alone, 20% for surgery alone, and 46% for combination therapy, demonstrating that the requirement for combination therapy reflected more severe cases.[7] Factors associated with a worse prognosis in the literature are as follows:

- Presence of bone destruction;
- Large primary tumor;
- Recurrence; and,
- Tumors located in the sphenoid sinus, maxillary sinus, or larynx.

All cases justify long-term monitoring because extramedullary plasmacytoma can recur as disseminated multiple myeloma and the evolution of this tumor is unpredictable.[1]

CONCLUSION

Extramedullary plasmacytoma is a rare, aggressive tumour that mainly affects the submucosa of the nasal cavity and paranasal sinuses. This tumour can remain in the area of the early lesion, advance to neighbouring areas, or even spread. The otorhinolaryngologist must identify the lesion and refer the patient for hematologic monitoring; moreover, a multidisciplinary approach is required to differentiate between localized disease and blood dyscrasias with a poor prognosis, such as multiple myeloma. The prognosis of extramedullary plasmacytoma is much more favourable than that of multiple

myeloma. Treatment with surgery followed by radiotherapy is effective. Controlled clinical trials are needed to establish a definitive treatment of choice for the management of these patients. The patient should always be monitored for a long period of time.

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