

Case Report

Extramedullary plasmacytoma nose operated by raveh's approach: a case report

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ABSTRACT

Plasmacytoma is a clonal proliferation of plasma cells. Plasmacytoma can be disseminated in the form of multiple myeloma or solitary disease. Most common site of involvement of solitary extramedullary plasmacytoma being the upper respiratory tract. We present a case report of 60-year-old male who presented with bilateral nasal obstruction, epistaxis and swelling on the root of nose. Biopsy proved out to be a plasmacytoma which was operated by Raveh's approach. Raveh's approach is a very useful approach for skull base tumors in the sphenoidal plane with minimal morbidity, no frontal lobe retraction and watertight closure of dura and excellent postoperative recovery.

Keywords: Extramedullary, Plasmacytoma, Subcranial Raveh's approach

INTRODUCTION

Plasmacytomas refer to a group of lymphoproliferative disorders characterized by monoclonal expansion of plasma cells that produce a single immunoglobulin molecule.¹ Multiple myeloma is the most common form of plasmacytoma. Plasmacytomas can also present in solitary form in bones called solitary plasmacytoma bone or in soft tissues as extramedullary plasmacytomas. It is believed that solitary plasmacytoma of bone is a precursor of disseminated disease multiple myeloma.

The spread of solitary bone plasmacytoma for myeloma usually occurs 3-5 years after the primary diagnosis. It is represented by a solitary osteolytic lesion without systemic manifestations of multiple myeloma, and therefore has a favorable prognosis after treatment.

Extramedullary plasmacytoma is a plasma cell tumor that forms in soft tissues such as the lymph nodes, skin and mucosa; thus, by definition, this tumor has no origin in the myeloid tissue of bone. It cannot be said that these plasmacytomas are also a precursor of multiple myeloma or not. Extramedullary plasmacytoma corresponds to less than 10% of all plasmacytic tumors, representing less than 1% of all head and neck tumors and less than 0.5% of tumors of the aerodigestive tract.²

80 to 90% of the extramedullary plasmacytoma cases are found in the head and neck; these tumors mainly occur in the respiratory tract, especially the submucosa of the nasal cavity, paranasal sinuses, nasopharynx, oropharynx, and larynx.³ When these tumors are present in nasal cavities, they have to be excluded from malignancy of nasal and paranasal sinuses.

CASE REPORT

A 60-year-old male presented to us with complaints of nasal obstruction for the last 5 years, initially unilateral (left) later progressed to right side also associated with on and off epistaxis. There was swelling on the root of nose since last two years (Figure 1).



Figure 1: Preoperative image showing the lesion.

There was no history of decreased vision or diplopia. There was a history of nasal surgery seven years back. There was no neck lymphadenopathy. Examination revealed swelling of about 6×5 cm present over the root of nose which was soft, non-tender, no fluctuant and overlying skin was normal. Anterior rhinoscopy was done which showed pale pinkish mass present in left nasal cavity which was bleeding on touch. Eye examination was normal. Rest of ENT examination was also within normal limits.

Contrast enhanced computed tomography of nose and paranasal sinuses was done which demonstrated large polypoidal expansile mass in the left nasal cavity superiorly; eroding the cribriform plate extending upto the anterior cranial fossa, bilateral frontal sinuses, abutting the brain parenchyma extradurally. Erosion of the nasal septum, medial wall of left orbit with extension to the left maxillary sinus and left orbital extra-conal space was seen (Figure 2).

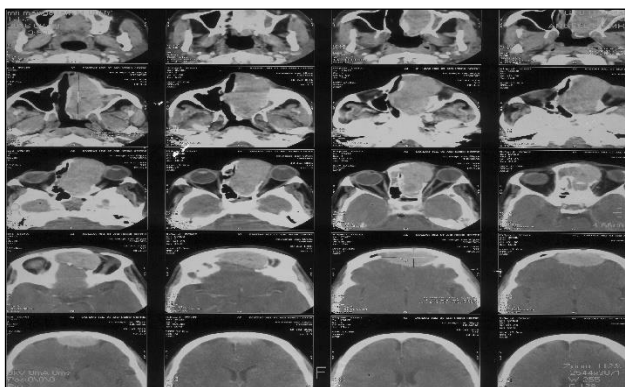


Figure 2: Radiological image (CECT) showing the lesion and the extent.

Biopsy of nasal mass was done under local anesthesia which showed plasmacytoma. Blood investigations were done to exclude multiple myeloma, blood calcium levels were normal. Kidney function tests are also normal. Bone marrow aspiration showed normal cellular reactive marrow, plasma cells were not increased. Final diagnosis of Sino nasal plasmacytoma was made. Excision of the mass was planned.

Subcranial raveh's approach was planned in which incision was made bilaterally from both the supraorbital notch medially, downwards along lateral nasal groove and over the dorsum of nose (Figure 3).



Figure 3: Intraoperative incision taken.

After elevation of flap tumor removal was done. Posterior wall of frontal sinus and lamina papyracea was eroded. Tumor attached to dura was removed by blunt dissection. Tumor going into left maxillary sinus was removed (Figure 4).



Figure 4: The defect after tumor removal.

Fascia lata and fat harvested from thigh and placed over the dura. Bilateral nasal packing was done. Packs were removed on the third postoperative day and oral feeding was started immediately postoperatively. Follow up showed no deterioration of vision and CSF leak. There was minimal postoperative morbidity. Patient had good aesthetic outcome (Figure 5).



Figure 5: Postoperative image of the patient.

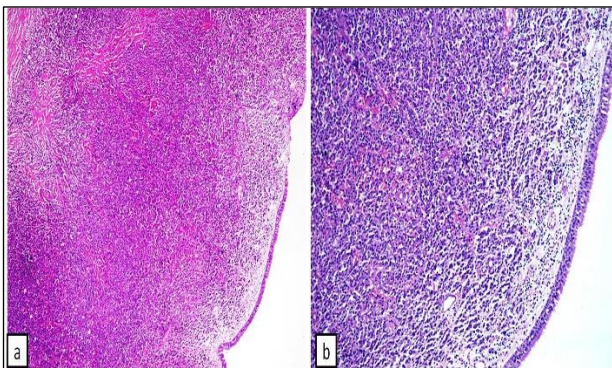


Figure 6a and 6b: (a) Respiratory epithelium with tumour in the submucosa, H&E stain (magnification 40x); (b) Tumor cells arranged in the form of sheets in the submucosa, H&E stain (magnification 100x).

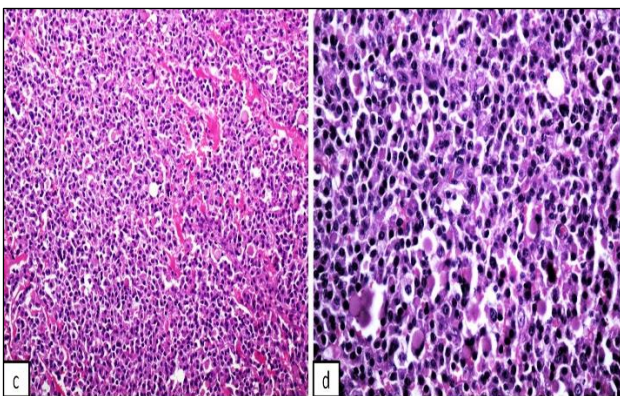


Figure 6c and 6d: (c) Sheets of plasma cells with few atypical plasma cells, H and E stain (magnification 200x), (d) Higher magnification showing sheets of plasma cells with few showing Russell bodies, H and E stain (magnification 400x); H and E: hematoxylin and eosin

Postoperatively histopathological analysis confirmed plasmacytoma. Microscopy showed partially denuded nasal mucosa with an uncircumscribed tumor in the nasal

submucosa. The tumor consisted of sheets of cells present singly. Cells were round to oval in shape with round to oval eccentric nucleus with cart-wheel chromatin. Cells had moderate amount of eosinophilic cytoplasm with some cells showing the characteristic Russell bodies seen in plasma cell neoplasms. Few atypical plasma cells showing high nucleo-cytoplasmic ratio with prominent nucleoli were also seen. Based on the above findings, a diagnosis of plasmacytoma of nose and paranasal sinus was given (Figure 6).

DISCUSSION

Extramedullary plasmacytoma is a rare, aggressive tumor that mainly affects the submucosa of the nasal cavity and paranasal sinuses. This extramedullary tumor was first described in 1905 by Schridde.⁴ The estimated global incidence of the disease is 1 case per 500,000 people.⁴ Over 80% of extramedullary tumors occur in the head and neck. Nose and paranasal sinuses is the most common site.³

Extramedullary plasmacytomas occur in people above 40 years of age (over 95% of cases) 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.

In the present case report patient is a 60-year-old male which is consistent with what is written in the literature. The clinical symptoms are more related to the specific location than to the nature of the tumor.

In a series of 20 patients with extramedullary plasmacytoma of the head and neck, Kapadia and colleagues observed the following major symptoms: tumor or local edema in 80%, nasal obstruction in 35%, epistaxis in 35%, localized pain in 20%, proptosis in 15%, rhinorrhea in 10%, regional lymphadenopathy in 10%, and paralysis of the VI cranial nerve in 5% of cases.⁵

When these tumors occupy the nasal cavities, a differential diagnosis should be made to exclude out other bleeding tumors, 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.

Wiltshaw stated in his study that 40% of extramedullary plasmacytomas spread beyond the primary site and/or undergo lymphatic drainage. Of these, 62% of patients had deposits in soft tissues and visceral organs, and 81% developed bone lesions.³ 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.⁶

The treatment of extramedullary plasmacytoma is controversial. According to Alexiou et al, sinonasal

EMPs with bone destruction should not be treated with radiotherapy or surgery alone, but rather with a combination of both modalities to assure local and systemic control.⁷ Galieni et al, reported that surgery in cases of limited and easily resectable masses would be adequate to treat the disease without recurrence.⁸

The approach to remove these extramedullary plasmacytomas is important and crucial for the complete excision. We used subcranial Raveh's approach for the excision.⁹ Various Craniofacial approaches have been described for most tumors, trauma, and congenital anomalies involving the anterior cranial fossa and the orbits, nasal cavity, or paranasal sinuses. The subcranial approach was pioneered by Raveh for repair of trauma, congenital anomalies and tumors of the anterior skull base. The subcranial approach differs from other orbitocranial approaches by including more of the nasal bones in the orbitonasal osteotomy and in not involving detachment of the medial canthal ligaments or mobilization of the orbits.

This approach provides broad anterior and inferior exposure of all the planes, including the anterior ethmoid roof up to the clivus, along with both orbital roofs towards temporal bone. This facilitates precise intradural and extradural tumour resection. The approach enables adequate visualization of tumour borders towards the dura as well as along the nasal and maxillary sinus extensions. The resulting simultaneous exposure of cranial as well as caudal aspects of the tumour, enables radical resection of tumour, while preserving the optic nerve, optic chiasm and carotid arteries, when these structures are not directly involved. It eliminates facial incisions and thus immediate postoperative edema and further facial deformities and also lead to decreased complication rates of Csf leak and infection.¹¹

This approach has been described in the literature as giving a bicoronal incision and elevation of subperiosteal flap preserving the pericranium. The flap is then dissected down to the frontozygomatic sutures bilaterally, and to the rhinion and piriform apertures in the midline. Subsequently, the periorbita is dissected from the superior, medial and lateral walls of the orbit, back to the apex of orbit on either side. Next, the outline of nasofrontal segments is planned, depending on the size and extent of both the lesion and frontal sinus. Before beginning osteotomy, the miniplates for subsequent bone fixation is adapted and drilled. The osteotomy lines are made across the frontal bone, down to and along the orbital roofs, down the medial orbital wall, and along the nasomaxillary grooves just anterior to the lacrimal duct. Subsequently, a vertical osteotomy performed anterior to the crista galli, allows detachment of the frontonasal segment, avoiding damage to the sagittal sinus or dural tears. In accordance with the location of tumour, two types of osteotomies are described - Type I: The first type involves an osteotomy of the frontonasal segment, leaving the posterior wall of frontal sinus intact, so as to

be removed in a second step. The procedure is indicated in cases where the tumour involves the posterior wall.¹²

Type II: This procedure involves single stage removal of the frontonasal segment, including the posterior wall of frontal sinus. The procedure is indicated for tumours not involving the posterior wall. The smaller dural defects may be sutured, but the larger defects resulting from intradural tumour involvement are patched with fascia lata.¹²

Therefore, Raveh's approach is a very useful approach for skull base tumors in the sphenothmoidal plane with minimal morbidity, no frontal lobe retraction with watertight closure of dura and excellent postoperative recovery.

CONCLUSION

Extramedullary plasmacytoma is a rare, aggressive tumor that mainly affects the submucosa of the nasal cavity and paranasal sinuses. A multidisciplinary approach is required to differentiate between localized disease and blood dyscrasias with a poor prognosis, such as multiple myeloma. Controlled clinical trials are needed to establish a definitive treatment of choice for the management of these patients. If surgery is planned adequate approach should be selected to remove the lesion with less complications and minimal morbidity. The patient should always be monitored for a long period of time.

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