Extramedullary Plasmacytoma of the Sphenoid Sinus presenting with visual loss: a Case Report

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Abstract

A rare case of sphenoid plasmacytoma in a 57-year old female who was presented with a frozen eye is reported. It was histopathologically confirmed based on transsphenoidal biopsy. Surgical removal of the tumor with complementary radiotherapy was performed as a treatment modality

Introduction:

Extramedullary plasmacytomas are localized plasma cell tumors that arise in tissues outside of the bone marrow which appear to be biologically distinct from solitary plasmacytoma of bone and plasma cell myeloma. Extramedullary plasmacytoma constitutes less than 5% of plasma cell neoplasms.[1,2]Extramedullary plasmacytoma is localized to the upper respiratory tract (nasal cavity and sinuses, nasopharynx and larynx) in over 80% of cases.[3]

In the sinonasal region, the clinical presentation includes nasal obstruction (29.8%), epistaxis, facial swelling, pain, rhinorrhea cranial nerve palsy, and visual loss .cervical lymphadenopathy presented in 5-20%of cases. [4]

According to the radiosensitivity of these tumors, radiotherapy remains the treatment modality of choice. Besides, surgery is suggested to obtain tissue for diagnosis, small localized lesions, and residual disease. [3]

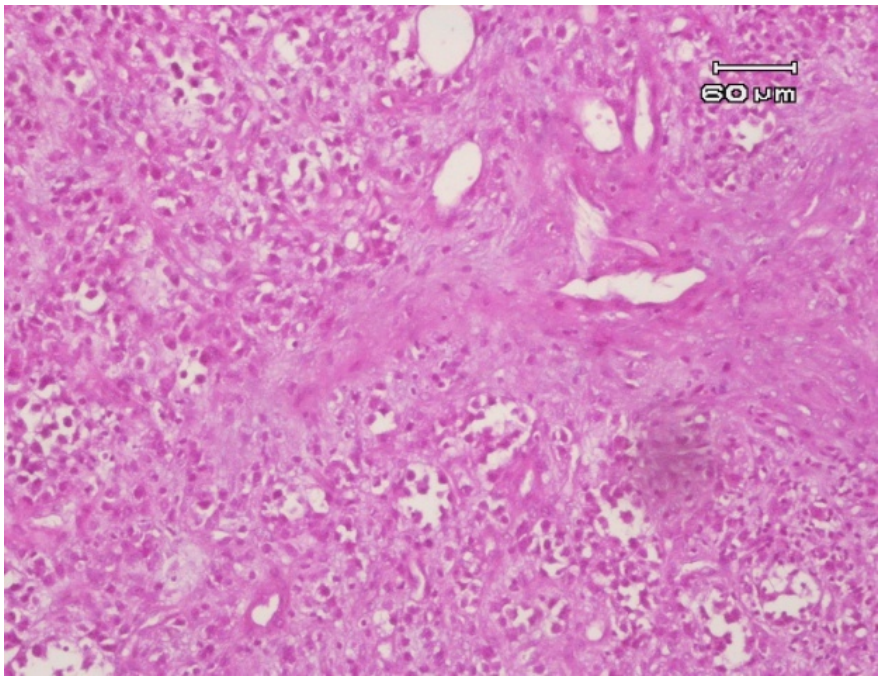
In this report, we describe a 62-year-old female with solitary extramedullary plasmacytoma arising from the right sphenoid sinus to highlight clinical and histological features.

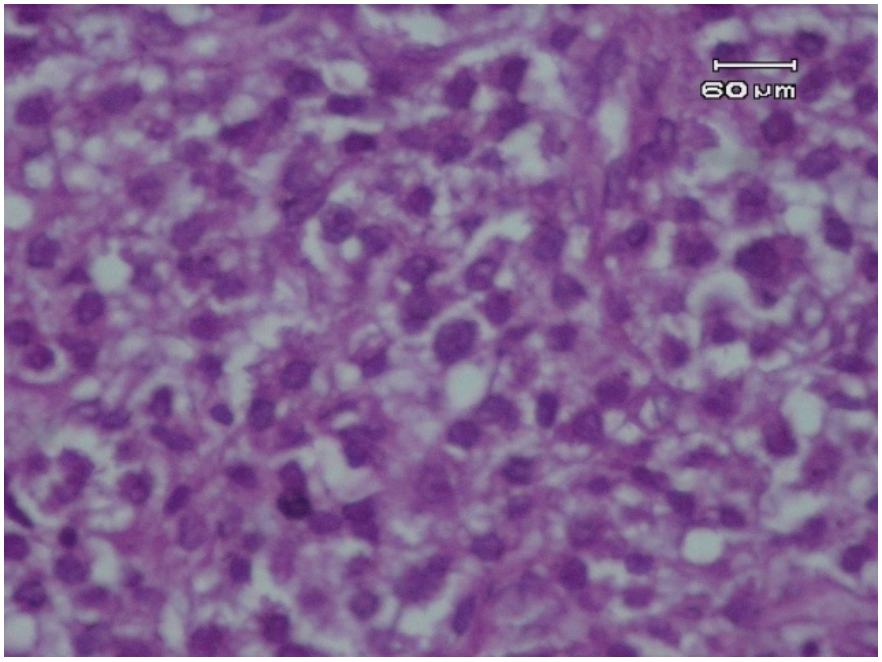
Case Report:

A 57-year-old female was referred to the department of otolaryngology and head and neck surgery, with 3 months history of ptosis and headache. On admission there was complete immobilization and decreased visual acuity of the right eye; the patient's past medical and drug history was unremarkable. There was no history of weakness, weight loss, nasal discharge, infectious diseases, or bone pain. On physical examination patient appeared well, and no lymphadenopathy was found. Abdominal examination showed no hepatomegaly or splenomegaly. A neurologic examination of cranial nerves revealed paralysis of right III, IV, and VI of cranial nerves and also hypesthesia of the right lateral face in the region of V1 and V2 of the trigeminal nerve.Furthermore, fundoscopy evaluation of the right eye revealed an atrophic optic disc; other neurological examination produced normal results.

Computerized tomography (CT) scan and magnetic resonance imaging (MRI) demonstrated an expansile mass lesion in the right sphenoid sinus eroding the sinus lateral wall and roof. The mass extension was observed in the right orbital apex and right superior orbital fissure and around the right internal carotid artery (figure2, 3).

Endonasal endoscopic observation and biopsies were performed under general anesthesia. Plasmacytoma was confirmed by histological analysis of multiple biopsy specimens of sphenoid sinus mass. Tumor samples were composed of several pieces of whitish-yellow soft tissue. The histopathological examination revealed monoclonal infiltration of plasma cells with atypical vesiculo nucleated nuclei and occasional binucleate forms; cytoplasm was basophilic in most cells (figure 1).





A B

figure 1A. monoclonal infiltration of plasma cells, B. with atypical nuclei and occasional binucleate forms and basophilic cytoplasms.

Immunohistochemistry was done for kappa and lambda light chains that was positive for lambda chains which confirmed the monoclonal nature of plasma cells.

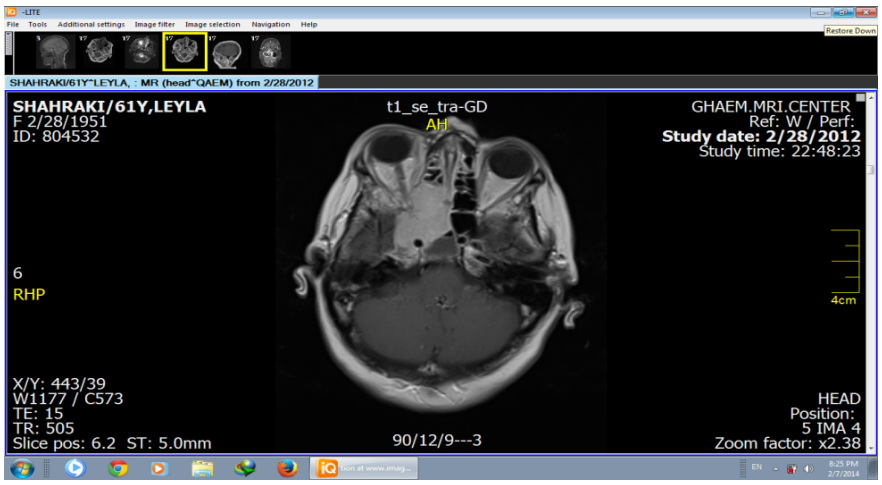
Further diagnostic work-up was performed. Complete blood cell count and biochemistry except slight anemia was all normal. The results of serum immunoglobulin and protein electrophoresis concentration and distribution were normal. (Table1) bone marrow aspiration and biopsy were normal and plasma cells represented <6% of all nucleated cells. Moreover, Test for serum myeloma protein and Bence-jones protein in urine was negative.Besides,no osteolytic lesions were identified in the skeletal survey.also,the abdominal ultrasonography was normal. These findings confirmed the diagnosis of extramedullary plasmacytoma of the sphenoid sinus.

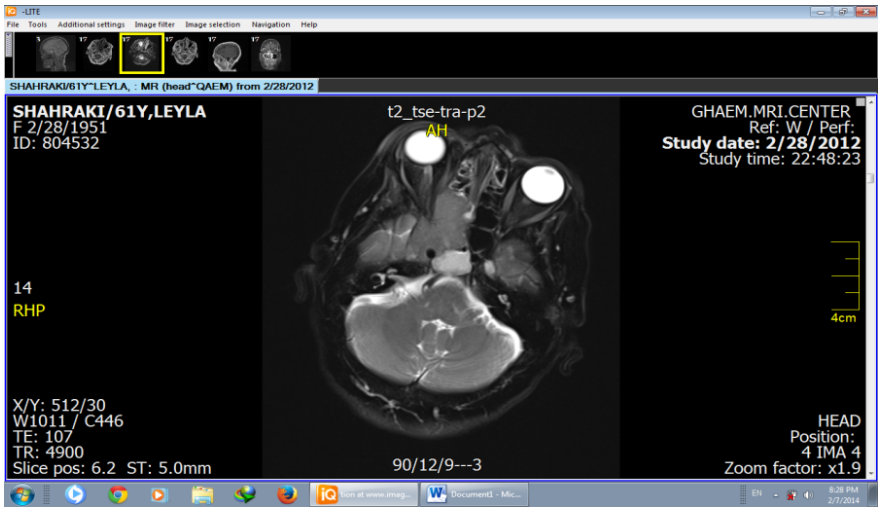
Surgical resection was performed with an endoscopic endonasal transsphenoidal approach by going through the opening of the left sphenoidal sinus and dissection of medial wall of right sphenoid sinus, the mucus layer of sphenoid sinus was removed and anterior wall of sphenoid sinus dissected and the tumor extracted through the inferior region of the sinus. Lateral extending of the tumor to the medial cranial fossa and surrounding optic nerve dissected and optic nerve decompressed; during the procedure, dura remained intact.

Table 1 results of serum immunoglobulin and protein electrophoresis

Value	Result	Reference range
B2-Microglobulin	4.072(mg/L)	1.22-2.46
Serum IgM	<0.247(g/L)	0.40-2.63
Serum IgG	10.78(g/L)	6.58-18.37
Serum IgA	<0.363(IU/ml)	0.71-3.60
Serum IgE	36.56(IU/ml)	Up to 182
Alpha1	3.90(4.5%)	0.21-0.35(29-4.9%)
Alpha2	0.74(9.2%)	0.51-0.85(7.1-11.8%)
Beta1	0.3(3.7%)	0.34-0.52(4.7-7.2%)
Beta2	0.23(2.9%)	0.23-0.47(3.2-6.5%)
Gamma	2.47(30.9%)	0.8-1.35(11.1-18.8%)

After surgical resection of tumor, complementary radiotherapy was performed. The patient is being observed closely during next 6 months. There have been some improvements in clinical symptoms of patient. paresis of ocular muscles somewhat resolved but decreased visual acuity still exists.





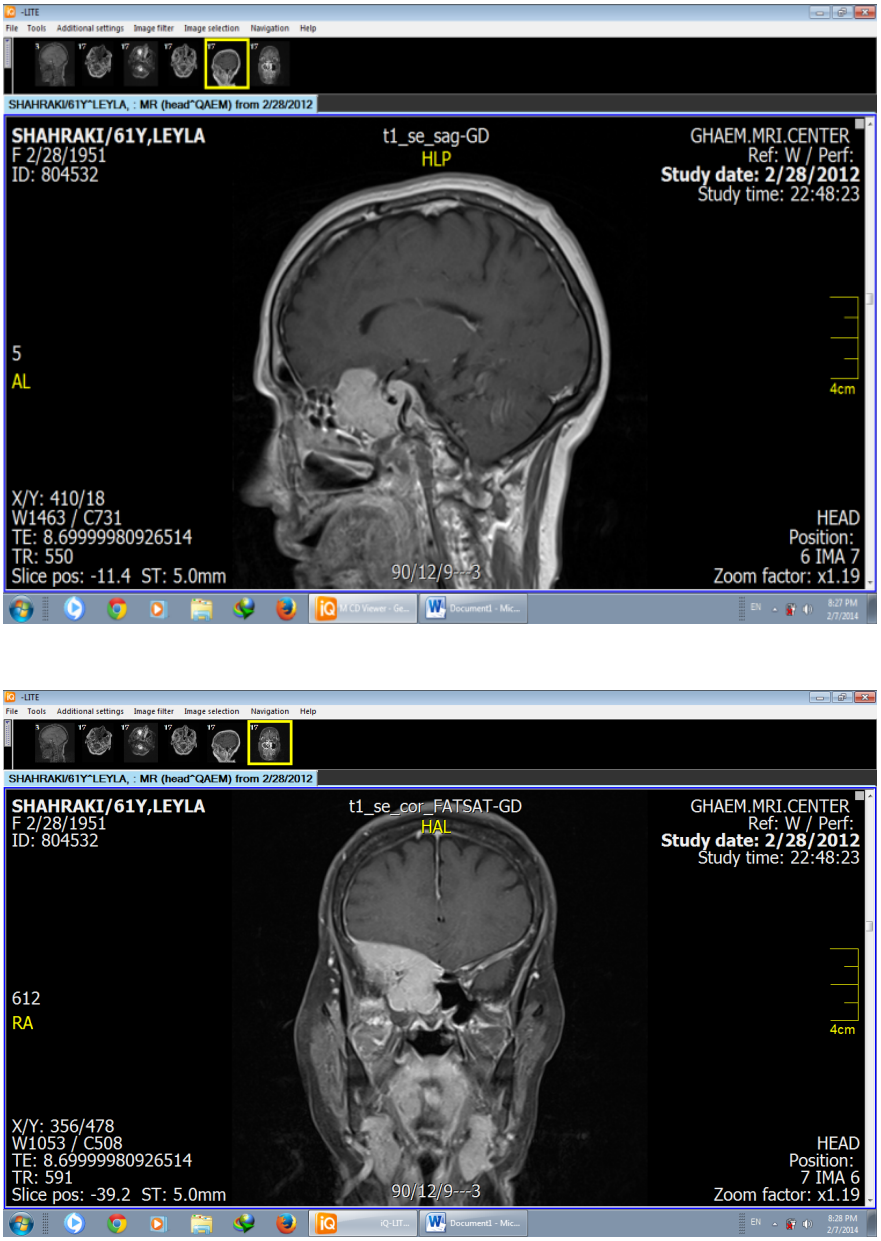
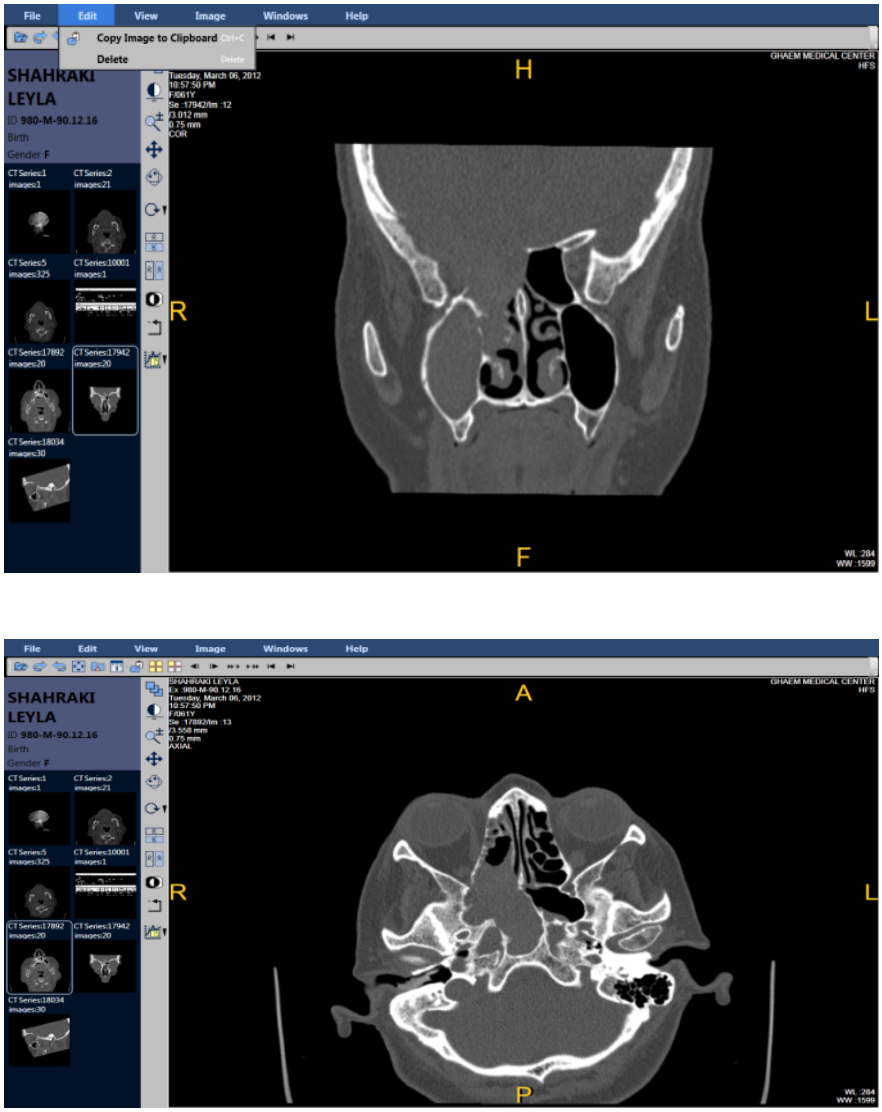


Figure2. Assessment of tumor extension. an expansile mass of right sphenoid sinus , eroding the bone laterally and superiorly, pressing against the optic nerve . The mass encase internal carotid artery, but does not cause stenosis (A and B) with homogenous enhancement (C and D).A and B: Axial T1W and T2W MR image; and C and D: sagital and coronal gadolinium-enhanced T1W MR images.



A B

Figure 3. CT-scan without contrast showing a homogeneous mass of the right sphenoid sinus eroding the sphenoid sinus lateral wall and roof, extending into the middle cranial fossa. A and B: coronal and axial image.

Discussion:

Plasmacytomas are immunoproliferative, monoclonal tumors of the β-cell line and are classified as non-Hodgkin's lymphoma. Extramedullary plasmacytomas present as a localized mass lesion. About 75% of them occur in the upper respiratory tract and also they may occur in a variety of anatomic sites [2,4]. The differential diagnoses of extramedullary plasmacytoma include reactive lesions with an abundance of plasma cells, lymphomas with marked plasma cell differentiation. Distinction from lymphoma with extreme plasma cell differentiation may be problematic[5]. Lymphoplasmacytic lymphoma, immunoblastic or plasmablastic large cell lymphomas, and especially MALT may be misdiagnosed as plasmacytoma[6,5]. Moreover, the distinction of extramedullary plasmacytoma from both solitary plasmacytoma of the bone and multiple myeloma is sometimes very difficult.

Mayoclinic diagnostic criteria for solitary plasmacytoma include 1) biopsy-proven solitary lesion of bone or soft tissue with evidence of clonal plasma cell,2) normal bone marrow with no evidence of clonal plasma cells, 3) normal skeletal survey,4) absence of end-organ damage such as anemia, hypercalcemia, renal failure or additional lytic bone lesions,5) low or absent serum or urinary level of monoclonal immunoglobulins[5].

In the present case, a systemic workup for finding dissemination of tumor cells including bone marrow aspiration, skeletal radiographic survey, complete blood count, serum biochemistry analysis, and monoclonal immunoglobulin levels of serum and urine did not reveal evidence of spread and the sphenoid sinus was the sole site of tumor. Therefore, extramedullary plasmacytoma of the sphenoid sinus was confirmed in this clinical case.

Based on the literature review, extramedullary plasmacytomas limited to the sphenoid sinus are extremely rare. It was estimated that only 1.6 % of extramedullary plasmacytomas arise from the sphenoid sinus that concluded approximately 15 cases until 2013. Moreover, in the localized sphenoid sinus tumor, the symptoms are usually non specific. when the tumor extended, visual loss, diplopia and, facial pain can be presented. [7, 8] Humphrey et al [9] reported an extramedullary plasmacytoma of the sphenoid sinus that presented with isolated VIth-nerve paralysis. In our presented case progressive ocular muscle paralysis and ptosis were the clinical manifestations. Orbital space-occupying lesions presenting with painful ophthalmoplegia are mostly due to malignancies that may originate from inside or outside of the globe [10]. In our case, the sphenoid originated tumor extended to the right orbital globe. Ampil et al [11] reported cavernous sinus involvement by extramedullary plasmacytoma of the sphenoid sinus which presented with retro-orbital headache. CT-angiography in our case showed cavernous sinus involvement and the patient‘s first complaint was headache 3 months earlier to admission.

The typical treatment for extramedullary plasmacytoma is local radiation, usually with radiotherapy in the range of 40 to 50 Gy[12]. Galieni et al [12] suggested that surgical removal of solitary extramedullary plasmacytoma could be performed for small masses and as secondary therapy after failure of local irradiation in the elimination of mass. Miller et al [13] reported a case of sphenoid sinus plasmacytoma that was treated with radiation 6400 cGy. Chemotherapy has not been successful in extramedullary plasmacytoma treatment but Wein et al [14] treated a patient with sphenoclival plasmacytoma with systemic chemotherapy. Surgical resection of tumor by endonasal transsphenoidal approach with complementary radiotherapy was performed for our patient. After 6 months of follow-up, there have been some improvements in the clinical symptoms of the patient. Although, paresis of ocular muscles is somewhat resolved, decreased visual acuity still exists. Hardwood et al [15] reported two cases of radiotherapy failure in the treatment of solitary extramedullary plasmacytoma which one of them was in the sphenoid sinus. With this in mind, 10-year disease-free survival rates are reported to be 70-80% [5], further follow-up with greater care is required for our patient.

Conclusion

Solitary extramedullary plasmacytoma of the sphenoid sinus is rare .In localized tumor; the symptoms are usually non specific. When the tumor extended, visual loss, diplopia and, facial pain can be presented. Radiotherapy remains the treatment modality of choice. Besides, surgery is suggested to obtain tissue for diagnosis, small localized lesions, and residual disease.

Acknowledgement

The authors would like to thank the patient for giving consent.

Data availability statement :

The data that support the findings of this study are available on request from the corresponding author, [Mahboobe Asadi].

CONFLICT OF INTEREST

The authors made no disclosures.

AUTHOR CONTRIBUTIONS

SS and MA contributed to the manuscript preparation and patient management. All authors read and approved the final manuscript.

ETHICAL APPROVAL

Because this report involves no experiment, ethics approval is waived.

Informed Consent .

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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