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CASE REPORT

Spheno-orbital Meningiomas: A Case Report

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Abstract

Spheno-orbital meningiomas are benign tumors. They arise intracranially from the sphenoid ridge arachnoid villi cap cells, extending into the orbit. Herein, the authors report a case of a 50-year-old female who presented to Ophthalmology department with a complaint of right eye proptosis. Magnetic resonance imaging and biopsy showed a right sphenoid-orbital meningioma. The tumor was excised and the patient symptomatically improved. Patient had gross tumor reduction for right sphenoidal orbital extra-axial brain tumor with hyperostosis of the lateral wall of the orbit and sphenoidal ridge.

Key words: Arachnoid, Brain neoplasms, Exophthalmos, Hyperostosis, Meningioma, Magnetic resonance imaging

Introduction

The term "meningioma" describes tumors that originate from the meningeal layer covering the brain and spinal cord. The most common orbital component arises from the superior orbital fissure, but optic canal extension is occasionally seen.

Spheno-orbital meningioma can have an intracranial and an intra-orbital component. The intracranial component arises from the anterior or middle cranial fossa. The intraorbital soft tissue component is associated with hyperostosis and bone involvement of the greater wing of the sphenoid bone. Meningiomas account for approximately 20% of all primary brain tumors, whereas autopsy reports are closer to approximately 30%.[1,2] The incidence of meningiomas increases with age, especially after age 65 years. It affects women more than men and African Americans more frequently than whites.[2-4]

Case Report

A 50-year-old female patient presented to the ophthalmology out-patient department on the 19th of November 2020. The patient complained of right eye progressive proptosis. The patient also complained of a subjective feeling of color desaturation since January 2020. The symptoms progressed more recently. No known medical illnesses were reported. The patient's past surgical history was insignificant expect for hysterectomy many years ago. The patient did not have any previous ocular surgery. Best corrected visual acuities were 6/6 in both eyes. Intraocular pressures were 15 mmHg in the right eye and 16 mmHg in the left eye. There wasn't a change in intraocular pressures upon up gaze. Restriction of extraocular motility in up gaze of the right eye was observed. Both eyes showed clear coronae. Anterior segments were normal in both eyes. The pupils were

round, regular, and reactive to light in both eyes. Relative afferent pupillary defect was not present. Central corneal thicknesses were 515 micrometers in the right eye and 514 micrometers in the left eye.

Patient was admitted for evaluation. Laboratory investigations were the following (White blood cells: 6.3 × 10^9/L; Hemoglobin: 11.4 g/dl; Platelets: 226 x 10^9/L; Erythrocyte sedimentation rate: 17 mm/hour; C-reactive protein: 4.03 mg/L; Fasting blood glucose: 5.6 mmol/L; Rheumatoid factor: 10 IU/ml; Thyroid stimulating hormone: 1 IU/ml; Anti thyroglobulin: 2 IU/ml; Anti-thyroid peroxidase: less than 28 IU/ml).

Blood investigations showed negative results for the following: Antinuclear antibodies (ANA); Cytoplasmic anti-neutrophil cytoplasmic antibodies (C-ANCA); Perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA), Human leukocyte antigen (HLA-B27), Anti-hepatitis C virus, Anti-Hepatitis A antibodies, Hepatitis B surface antigen. The patient's complement factor levels were C3 149 mg/dl and C4 32.5 mg/dl. Magnetic resonance imaging (MRI) of her brain and orbit revealed hyperostosis of the greater wing of right sphenoidal bone. Right lateral orbital wall and orbital roof expansion by well-defined lesion of dark signal on all pulse sequences measuring 4.3 x 3.3 x 3.5 centimeters, indenting the posterior and lateral orbital wall. The lesion caused right lateral rectus muscle displacement and right eye mild proptosis. Mild indentation on right optic nerve was noticed (Figure 1). Findings were compared with computed tomography (CT) findings which showed diffuse sclerosis and hyperostosis with bony expansion involving right sphenoid wing.

Patient was re-evaluated when she was admitted under neurosurgery care on the 24th of December 2020. The patient complained of upper lid swelling with color desaturation and decreased contrast sensitivity. The patient experienced limitation in up gaze and dextro-version. The patient also complained of diplopia in up gaze, dextro-version, and inferior gaze. Hertel ophthalmometer readings were 26 millimeters in right eye and 20 millimeters in left eye. Best corrected visual acuities were 6/6 in both eyes. Intraocular pressures were 20 mmHg in right eye and 12 mmHg in left eye.

Patient was taken for gross tumor reduction (GTR) under general anesthesia. Histopathological specimen showed meningothelial meningioma. The tumor was classified by World health organization (WHO) as grade 1, with positive immunohistochemical stain for epithelial membrane antigen (EMA). The patient's symptoms improved, and proptosis subsided in follow up appointment. Hertel ophthalmometer readings were 22 millimeters in right eye and 20 millimeters in left eye. Post-operative MRI images after the patient underwent GTR are shown in Figure 2. There was a significant reduction in the right eye proptosis and improvement of the lateral rectus muscle position. Best corrected visual acuities were 6/6 in both eyes. Radiation therapy was not done. Patient had regular follow up appointments with the ophthalmology department.

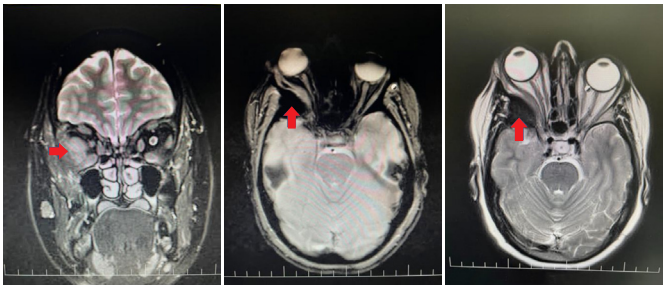

Figure 1: "Mass-like" sphenoid-orbital meningioma with hyperostosis in the greater wing of right sphenoidal bone. Right lateral orbital wall and orbital roof expansion by well-defined lesion of dark signal on all pulse sequences measuring 4.3 x 3.3 x3.5 centimeters. There is indentation of the posterior and lateral orbital wall and displacement of right lateral rectus muscle causing a mild right eye proptosis. Mild indentation on right optic nerve was noticed

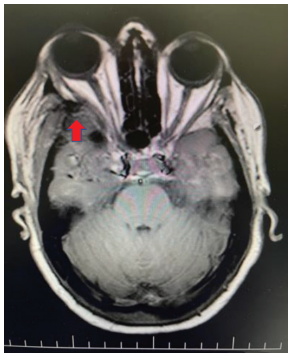

Figure 2: Demonstrates post-operative MRI images after the patient underwent gross tumor reduction. A significant reduction in the right eye proptosis and improvement of the lateral rectus muscle position

Discussion

Sphenoidal orbital extra-axial brain tumor, with hyperostosis of the lateral wall of the orbit and sphenoidal ridge, can be surgically excised to improve visual function and proptosis. In cases of symptomatic or progressively enlarging tumors, complete surgical excision, surrounding dural attachment and involved bone is recommended.[4] If complete excision is not possible, other options including definitive external-beam radiation and partial excision followed by adjuvant radiotherapy are considered. Long-term data using definitive external-beam radiation have demonstrated prolonged tumor control comparable with that observed with surgery followed by adjuvant radiation.[5] In this case external-beam radiation was not indicated because surgical excision was sufficient to debulk majority of the tumor.

This case has a 10-year risk recurrence rate of 9% according to the Simpson grades of resection, as derived from 265 patients.[6] Meningiomas rarely metastasize with approximately 0.001% of distant extracranial metastases, most commonly to the lung.[7,8]

The provisional diagnosis of orbital proptosis includes thyroid disease related proptosis, idiopathic orbital Inflammation, metastasis, orbital lymphoma, and meningoencefalocele.[9] This patient had normal thyroid function tests; therefore, thyroid eye disease can be ruled out in this case. Moreover, primary orbital lymphomas can be ruled out by the histopathological results. Primary orbital lymphomas are very rare tumors.[10] They are mostly of B cell origin.[11] Orbital hematoma was ruled out because there wasn't any history of trauma or anticoagulation medication use and the proptosis was longstanding. Orbital meningiomas can be confused with idiopathic orbital inflammation. Rodríguez-Colón reported a case of sphenoidal meningioma mimicking idiopathic orbital inflammation.[12] This patient showed normal inflammatory markers which make idiopathic orbital inflammation highly unlikely. In addition, patient did not show any leukocytosis and did not have a history of fever, therefore, causes from an infectious origin can be excluded from the differential diagnosis. Meningoencephalocele was excluded by MRI images. Unlike meningioma, meningoencephalocele presents as a combination of fluid-filled cyst and parenchyma with no contrast enhancement and shows a continuity between brain parenchyma and herniated meningoencephalocele on MRI images.[9]

The patient was diagnosed with Spheno-orbital meningioma through specific findings in the MRI images and specimen biopsy of the lesion which specifically stained positive for EMA. The patient underwent gross tumor reduction by neurosurgical and orbital intervention. Sphenoid-orbital meningiomas requires a multidisciplinary surgical approach.[13] The patient's signs and symptoms improved and has a low risk of recurrence because biopsy showed a WHO tumor grade 1. The WHO tumor grade and tumor marker can predict the behavior of the meningioma and the risk recurrence.[14] Kiyofuji described aggressive removal of involved bone and periorbita is crucial. Proptosis and visual field defect other than a central scotoma can improve after surgery.[15]

Conclusion

Spheno-orbital meningiomas are benign tumors. They arise intracranially from the sphenoidal bone. This patient was found to have right sphenoidal orbital extra-axial brain tumor, with hyperostosis of the lateral wall of the orbit leading to the diagnosis of sphenoid-orbital meningioma. Patient's symptoms improved after she underwent gross tumor reduction.

Disclosure of interest

The authors declare that they have no competing interest.

Patient consent

Written consent was obtained from patient for the publication of this case report.

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