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Clinical Presentation and Management of 10 Consecutive Cases of Primary Orbital Tumors of Neurological Origin Presenting to a Regional Institute of Ophthalmology

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Abstract

Introduction: Proptosis is a common presentation to our outpatient department. The eyes are as much a visual structure as they are an aesthetic part of the face. We designed a study to document the clinical presentation and management of ten consecutive cases of primary benign neural orbital tumors presenting to a regional institute of ophthalmology.

Aim: The aim of the study is to document the clinical presentation and management of ten consecutive cases of primary benign neural orbital tumors presenting to a regional institute of Ophthalmology.

Methodology: Ten consecutive cases of primary benign neural orbital tumors were enrolled. They were subjected to a thorough clinical examination and imaging. Thereafter they were managed by observation, surgery or radiotherapy as per the case.

Results: There were five cases of schwannoma which responded well to surgery with good visual outcome. The two cases of optic nerve glioma one was subjected to observation due to slow growth and good vision. The second case was managed surgically due to proptosis and poor vision. Two cases of meningioma were subjected to surgery due to proptosis with poor vision. One case was observed and subjected to radiotherapy due to good vision and slow growth.

Discussion: We report the clinical presentation and management of primary orbital tumors of neurological origin from our part of the world. The current management scenario from our perspective is in concordance with that reported by other workers from different parts of the globe.

Conclusion: Primary benign neural tumors of the orbit can be managed by surgery or observation as per the case. Our unique perspective on these tumors is invaluable to the world oncology literature.

Keywords: Orbital neurlogical tumors; Management

Introduction

The ability to see is a divine gift. The ability to see our own selves as healthy and beautiful is another gift. It would be distressing to lose both vision and cosmesis attributable to a tumor behind the eyeball.

Orbital tumors of neurological origin represent a distinct clinical group presenting to our regional institute of ophthalmology. We are aware of optic nerve tumors for two centuries since Antonio Scarpa first clinically reported optic nerve tumor in 1816 [1].

Medical science has evolved by leaps and bounds over the centuries. We designed a study to document the current management approach of these tumors at our regional institute of ophthalmology.

The study showcases the clinical presentation and current management of benign primary orbital neural tumors. We report the current clinical scenario from our unique perspective. Our clinical experience in this part of the globe adds valuable insights to the current management options of these diverse orbital tumors.

We present a case series of ten cases seen over a period of seven years.

Aim

The aim of the study is to document the clinical presentation and management of ten consecutive cases of primary benign neural orbital tumors presenting to a regional institute of Ophthalmology.

Methodology

The study was carried out at M & J Western Regional Institute of Ophthalmology, Ahmedabad from February 2011 to February 2018 (seven years). Ten cases of orbital tumors of neurological origin were enrolled. The patients were followed for a period of one year.

The study was ethically approved by the institutional review board. Case enrolment included cases of proptosis, all age groups, both the

genders presenting to the institute's outpatient department. All cases diagnosed as primary neural benign orbital tumors on basis of clinical presentation and imaging were enrolled. Clinical presentation was insidious onset, chronic axial proptosis. Diminution of vision, relative afferent pupillary defect, reduced colour vision, disc swelling, optociliary shunt vessels were helpful in pointing the diagnosis towards a neurological tumor. Blood investigations ruled out an inflammatory cause.

A computed tomography scan preferably with contrast enhancement was carried out of both orbits including brain and paranasal sinuses. The advances in imaging techniques have made it possible for us to know the exact location of the lesion. We can also accurately predict the histological nature of the lesion *via* imaging alone which helps us to plan management of these lesions scientifically.

Optic nerve glioma appeared on imaging as a spindle shaped tumor arising from the optic nerve, the optic nerve not visualized separately from the nerve. There is no calcification and no contrast enhancement.

Meningioma of the optic nerve sheath appears as an intraconal mass with indistinct margins, contrast enhancement, calcification and tram track appearance. On coronal view the tumor was visualized as a fluffy mass with indistinct borders. A hypodense spot was visualised in the centre of the lesion which was the optic nerve.

Schwannoma presented as an intraconal soft tissue mass with distinct margins, minimal contrast enhancement and bony expansion.

Results

We report ten cases of primary neural orbital tumor.

There were five cases of schwannoma which responded well to surgery with good visual outcome. The patients were in the age group of 25-40 years, 2 males and 3 females. The clinical presentation was painless proptosis. On histopathology Schwannoma appeared as densely packed spindle shaped cells with pallisaded nuclei.

There were two cases of optic nerve glioma .The patients were of the age of 7 years and 12 years respectively. They presented with painless proptosis of one and half year and one year duration respectively. First patient was subjected to observation due to slow growth and good vision. Radiotherapy was not considered as there was no chiasmal involvement or rapid growth. The second case was managed surgically due to proptosis and poor vision. On histopathology they appeared as pilocytic astrocytomas.

There were three cases of optic nerve meningioma. Two patients ladies, one thirty two years and the other twenty five years were subjected to surgery due to proptosis with poor vision. On histopathology psammoma bodies were seen as oval to polyhedral cells with vesicular nuclei and acidophilic cytoplasm arranged in nests and whorls. There was no recurrence at two year follow up in both patients.

One case of meningioma in a lady aged thirty eight years was observed due to good vision and slow growth. The case was referred to a radiation oncologist where radiotherapy in fractionated doses was given. On our follow up at one year the clinical presentation and imaging indicated that the lesion had stabilized.

Discussion

The plethora of orbital diseases makes it imperative for us to segregate patients for optimum management. The inability to directly visualize and palpate the orbital space occupying lesions makes it a challenge. The constellation of symptoms and leading signs are a clue to the definitive diagnosis. The imaging and histopathological aids are invaluable.

We managed our cases with surgery or observation/ radiation as per the case and available resources.

Knapp et al. documented the management of optic nerve glioma as observation for patients with isolated optic nerve involvement and good vision. Patients must be reviewed regularly with repeat serial MRI scans of the optic nerves [1]. They recommend that lesions involving the chiasma or the optic tracts must be considered for radiotherapy or chemotherapy.

Knapp et al. documented the management of optic nerve meningioma as surgery or radiotherapy. Surgery may be associated with recurrences or orbital invasion. Radiotherapy has its set of side effects. Observation alone may also be an option for extremely slow growing tumors [1].

Knapp et al. reported the management of orbital schwannoma as surgery [1].

Rootman and Robertson documented the management of schwannoma (neurilemmoma) as slow growing, solitary, noninfiltrative, benign mass which can be removed surgically successfully [2].

Rootman and Robertson documented the management of optic nerve meningiomas as observation alone. Therapeutic interventions can be in the form of radiotherapy or surgery. This is indicated when there is evidence of or risk of spread to the central nervous system, progressive or aggressive growth and visual deterioration [2].

Rootman and Robertson documented the management of orbital optic nerve glioma as observation only. Surgical excision is indicated only if there is disfiguring proptosis or rapid progression of the tumor [2].

Dutton document management of optic nerve sheath meningioma as observation/surgery. He reports management of optic nerve glioma as observation/surgery. The management of Schwannoma is surgical excision [3].

Delfini et al. reported that patients with neurinoma of the orbital cavity have the most favourable prognosis in terms of visual function as well as long term quality of life. Removal of optic nerve glioma and optic nerve meningioma are associated with poor visual outcome [4].

We present the management of these cases from our perspective which is in keeping with the current management options of these tumors.

Conclusion

We report the clinical presentation and management of primary orbital tumors of neurological origin from our part of the world. The current management scenario from our perspective is in concordance with that reported by other workers from different parts of the globe.

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