Orbital Paraganglioma: A Systematic Review

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Abstract

Purpose The purpose of this study was to perform a systematic review of the literature on orbital paragangliomas, evaluating clinical presentation, diagnosis, management patterns, and prognosis.

Methods The systematic review was conducted based on the principles described in the Preferred Reporting Items for Systematic Reviews and Meta-Analysis. PubMed, Cochrane databases, Embase, and Web of Science were searched for articles related to orbital paragangliomas. Inclusion criteria included English language articles with original reports on human subjects. Data on clinical presentation, diagnosis, treatment, and prognosis were collected.

Results Twenty-seven articles met inclusion criteria (28 total patients). The mean patient age was 37.1 years (range, 3–75 years); 13 (46.4%) patients were male. The most common presenting symptoms were proptosis (89.2%), visual acuity changes (67.9%), and extraocular muscle restriction (64.2%). Lesions were most commonly intraconal (92.9%). Single modality therapy was employed in 19 patients (67.9%), including excision in 12 patients (42.9%) and exenteration in 5 patients (17.9%). Ten patients (35.7%) developed recurrence, and there was a 92.9% survival rate (mean follow-up, 29 months).

Conclusion Orbital paragangliomas are rare tumors with an excellent prognosis. These lesions commonly present with proptosis, and are primarily managed surgically with simple excision, although exenteration and adjuvant radiation may be necessary for invasive tumors. This series is the largest and most comprehensive systematic review of orbital paragangliomas conducted to date.

Keywords

- ▶ orbital paraganglioma
- ▶ systematic review
- ▶ PRISMA

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Introduction

Orbital paragangliomas are rare lesions first described in 1952.¹ Paragangliomas are growths of nonchromaffin paraganglionic tissue and can occur in the abdomen, thorax, or head and neck. In the head and neck region, they are commonly found associated with carotid bodies and seldom present in the orbit. Orbital paragangliomas are thought to originate from the ciliary ganglion.² The literature on orbital paragangliomas comprised exclusively of case reports; consequently, the clinical information available on this topic is sparse, and providing patients with prognostic information is a difficult task. The aim of this systematic review is to provide clinicians with detailed information on the epidemiology, treatment patterns, and prognosis of orbital paragangliomas. To our knowledge, this report is the only comprehensive systematic review to focus on orbital paragangliomas.

Methods

The systematic review was performed based on the principles set forth in the Preferred Reporting Items for Systematic Reviews and Meta-Analysis (►Fig. 1). PubMed, Web of Science, Embase, and Cochrane databases were queried for articles related to orbital paraganglioma using the following search terms: orbital paraganglioma, paraganglioma of the orbit, orbital chemodectoma, and orbital glomus body tumor. The search yielded 156 articles. The title and abstract were reviewed for all studies, pertinent articles were downloaded, and the references were reviewed to ensure all relevant information were included. Only studies that had individual patient information were included. Reports that were in a foreign language, contained insufficient patient information, or were about nonhuman subjects were excluded. Demographic information, clinical presentation, and treatment information were extracted from the articles.

Fig. 1 Article selection process based on Preferred Reporting Items for Systematic Reviews and Meta-Analysis.

Results

The literature search retrieved 156 articles. No other systematic reviews or meta-analyses were identified. Twenty-seven articles met inclusion criteria, 26 studies were single patient case reports, and 1 study contained two patients for a total of 28 unique patients.^{1–27}

Patient Characteristics

The mean patient age was 37.1 years (range, 3–75 years); 13 (46.4%) patients were male and 15 (53.6%) were female. The right eye was affected in 16 patients (57%). The most common presenting symptoms were proptosis (89.2%), visual acuity changes (67.9%), extraocular muscle restriction (64.2%), diplopia (39.3%), and eye pain (28.6%). Mean duration of symptoms at the time of initial presentation was 14.1 months (range, 2–48 months). Seventeen patients (60.7%) had postoperative complications, including loss of vision in 12 (42.9%) patients secondary to exenteration or optic nerve damage intraoperatively. Two patients (7.1%) died from the disease. Mean follow-up time was 29.4 months (range, 3–120 months) (►Table 1).

Table 1 Summary of patient characteristics

Category	Subcategory	Total
No. of patients		28
Average age at diagnosis (y)		37.1
Sex (M:F)		13:15
Length of symptoms (mo)		14.1
Laterality (R:L)		16:12
Clinical features		
	Proptosis	25 (89.3%)
	Visual acuity changes	19 (67.9%)
	Extraocular muscle movement restriction	18 (64.2%)
	Diplopia	11 (39.3%)
	Eye pain	8 (28.6%)
	Fundoscopic changes	15 (53.6%)
	Other symptoms	19 (67.9%)
	Sympathetic changes	1 (3.6%)
Local invasion		15 (53.6%)
Metastasis		3 (10.7%)
Loss of vision		12 (42.9%)
Recurrence		10 (35.7%)
Complications		17 (60.7%)
Mean Length of follow-up (mo)		29.4
Death from disease		2 (7.1%)

Table 2 Location of paragangliomas

Location of pathology	Total
Lateral orbital	6 (21.4%)
Medial orbital	9 (32.1%)
Inferior orbital	9 (32.1%)
Superior orbital	9 (32.1%)
Intraconal	26 (92.9%)
Intracranial	1 (3.6%)
Intraocular	1 (3.6%)
Attachment to optic nerve	8 (28.6%)

Tumor Characteristics and Diagnosis

The tumor was most commonly located intraconally (92.9%) and attachment to the optic nerve was noted in 8 (28.6%) patients. One case had intracranial involvement (►Table 2). Local invasion was reported in 15 (53.6%) patients, mostly involving the extraocular muscles. Metastasis occurred in three patients (10.7%) to lymph nodes (two) or to the vertebra (one). Local tumor recurrence occurred in 10 patients (35.7%). The size of the tumor measured from surgical specimen was reported in 13 (46.4%) cases (range, 2 cm × 1 cm × 1 cm–4.5 cm × 5 cm × 7 cm). Computed tomography (CT) was the most commonly used imaging modality for initial diagnosis (46.4%), and the use of magnetic resonance imaging (MRI) and radiography was used equally in 21.4% of cases, with MRI being used more often in the past decade. X-rays were predominantly used in developing countries. Radiography, however, was almost always inconclusive or negative. The confirmatory laboratory tests were electron microscopy (EM) studies and immunohistochemical staining (IHCS). Identification of membrane-bound electron-dense neurosecretory granules on EM is diagnostic, which was present in 100% of reported cases that used this imaging modality for diagnosis. With reported IHCS, positive reactions were seen with chromogranin (72.7%), synaptophysin (100%), neuron-specific enolase (75%), S100 (77.8%), and none with chromaffin (►Table 3).

Table 3 Electron microscopy and immunohistochemistry characteristics of paragangliomas

	Positive	Negative	Sensitivity (%)
Chromogranin	8	3	72.7
Synaptophysin	9	0	100
Neuron-specific enolase	6	2	75
Chromaffin	0	5	0
S100	7	2	77.8
Neuron-like cells on electron microscopy	8	0	100

Treatment

Orbital paragangliomas are most commonly managed surgically, including either simple excision or exenteration with adjuvant radiation used for more aggressive disease (►Table 4). Single modality therapy was used in 19 patients (67.9%), including excision 12 (42.9%) and exenteration 5 (17.9%). In other cases, either radiation therapy 1 (3.6%) or biopsy 1 (3.6%) alone caused the tumor to regress completely. No tumor recurrence was noted in patients treated with expectant management 1 (3.6%), biopsy alone 1 (3.6%), or excision combined with neurosurgical intervention 1 (3.6%). Recurrence rate varied with treatment. Tumor recurred in 16.7% of patients treated with excision alone (12) and in 40% of patients who were treated with exenteration alone (5). Two patients expired as a result of their disease. Chemotherapy was not used in any of the reported cases.

Patients with Visual Loss

Visual function is an important assessment of treatment outcome. Visual loss is defined as decreased visual acuity from baseline prior to the onset of orbital symptoms. Twelve (42.9%) patients reported visual loss and their characteristics are summarized in ►Table 5. The mean age of the patients was 34.9 years. Five of the patients (41.7%) had tumors attached to the optic nerve. Ten (83.3%) patients presented with visual disturbance at their initial clinical visit. Tumor recurrence rate in this group of patients was 58.3% and their mean follow-up time was 28.7 months.

Recurrence

Ten of 28 patients (35.7%) had tumor recurrence, and their characteristics are presented in ►Table 6. The mean patient age was 34.7 years. The tumor was attached to the optic nerve in 40% of patients. Two cases of mortality occurred in patients who had a recurrence. The survival rate was 80% and the mean length of follow-up was 29.6 months (►Table 6).

Discussion

Since Fisher and Hazard described the first case of orbital paraganglioma in 1952, there has been fewer than 50 cases reported worldwide.¹ The origin of the tumor has yet to be identified because the existence of paraganglionic chemoreceptors in the human orbit is still controversial. However, as demonstrated in case reports reviewed in this study, the occurrence of primary orbital paragangliomas with no coexisting extra-adrenal paragangliomas elsewhere supports the notion that paraganglionic chemoreceptors do exist in the orbit with the consensus being that the tumor arises from the ciliary ganglion.² Orbital paragangliomas can present in patients of all ages, affecting both genders equally and the most common location was intraconal. Although the tumor is benign in most cases, it may cause proptosis, extraocular movement restriction, diplopia, decreased visual acuity, and less frequently, eye pain (►Table 1).

Our results suggest that orbital paraganglioma is a benign tumor predominantly, with a recurrence rate in the literature of 35.7% and mortality rate of 7.1%. There are no histologic

Table 4 Summary of treatments performed

Treatment	Number of patient	Patients (%)	Age (y)	Gender (female, %)	f/u (mo)	Recurrence (%)	Loss of vision (%)	Survival (%)
Excision alone	12	42.9	37.9	66.7	27.1	16.7	8.3	100
Exenteration alone	5	17.9	35.2	40	39.8	40	100	100
Radiation alone	1	3.6	24	0	24	100	100	100
Biopsy alone	1	3.6	75	100	24	0	0	100
Excision + exenteration	1	3.6	13	100	5	100	100	100
Excision + exenteration + radiation	1	3.6	54	0	12	100	100	100
Exenteration + radiation	1	3.6	35	0	48	100	100	0
Excision + neurosurgery	2	7.1	52.5	100	4.5	0	0	100
Excision + radiation	2	7.1	35.5	0	34.7	50	100	50
Excision + neurosurgery+ radiation	1	3.6	31	100	48	100	0	100
No treatment	1	3.6	47	0	0	0	0	100

Abbreviation: f/u, follow-up.

criteria that reliably predict malignant transformation, hence it is defined by distant metastasis which was present in 3 (10.71%) of patients in our review. In 53.6% of patients, the tumor exhibited local invasion, most commonly into the extraocular muscles, resulting in decreased ocular motility and in some cases affecting visual acuity. Tumors with local invasion place the patients at increased risk of diplopia postoperatively.

Diagnosis

CT scan and MRI are both effective initial imaging modalities for localization and size assessment, although CT is used more frequently. The tumor appears as a well circumscribed, homogenous, and hyperdense lesion on CT scan, frequently associated with proptosis or displacement of the orbital content.¹⁸ On MRI, the tumor is well circumscribed with a “salt and pepper” appearance with both hyperdense and hypodense areas, which is pathognomonic for orbital paragangliomas. MRI is critical when there is concern for intracranial tumor extension to characterize tumor extent.²¹ IHCS and EM are the gold standard diagnostic tests for orbital paragangliomas. Synaptophysin was the most sensitive test with 100% sensitivity; neuron-specific enolase was second at 75% (► **Table 3**). Genetic analyses are a critical part of the patient work-up and analyses have identified six germline mutations that contribute to paraganglioma syndromes, with three mutations being related to the succinate dehydrogenase (*SDH*) gene.⁵ *SDH* is an enzyme in the mitochondria that plays a role in the citric acid cycle in oxidative metabolism. Patients with the *SDH* mutation tend to have a better prognosis and lack of *SDH* staining is an indicator of adverse prognosis.²⁸

Treatment

The primary treatment for orbital paraganglioma is surgical; 42.9% of patients received excision alone and 17.9% received

an orbital exenteration. In the United States, excision was more likely to be the initial intervention, whereas exenteration was the primary surgical option in developing countries, possibly due to technical requirements and limitations in resources.¹⁵ If the lesion is well encapsulated, the primary management should be excision through an orbitotomy.¹² Incomplete excision with close surveillance is an option for patients with orbital paragangliomas abutting the optic nerve (► **Table 4**).¹⁷ Of the eight patients in our study who had paragangliomas attached to the optic nerve, four (50%) developed recurrences. Exenteration should be offered to patients with rapidly expanding tumors, tumors resulting in a significant decrease in vision, tumors that are locally invasive resulting in cranial nerve neuropathies and for recurrent tumors.²⁹ Preoperative imaging is critical for surgical planning to determine ease of resection and feeding vessels. Preoperative embolization should be considered in patients with large tumor with distinct feeder vessels.³⁰ Radiation therapy should be reserved for patients who are not good surgical candidates, for patients with locally destructive disease where complete surgical resection is difficult or can compromise critical structures and as neoadjuvant therapy to arrest the growth of a rapidly growing tumor.¹⁷ In our series, five (17.9%) patients received radiation therapy, and only one received radiation alone.

Permanent Visual Loss

Patients who had permanent loss of visual acuity were more likely to have vision changes at initial presentation (83.3% compared with 67.9% for all patients). In addition, patients with permanent decrease in vision had an increased rate of tumor attachment to the optic nerve (41.7% compared with 28.6% in the general population studied). Hence, it is important to counsel patients who have tumors in close proximity to the optic nerve about the risk of a permanent decrease in visual acuity.

Table 5 Summary of patients with permanent vision loss

Patient characteristics	Subcategory	Totals
No. of patients with permanent visual loss		12 (42.9%)
Age (y)		34.9
Sex (M:F)		8:4
Clinical features		
	Proptosis	11 (91.7%)
	EOM restriction	10 (83.3%)
	Visual acuity changes	10 (83.3%)
	Eye pain	5 (41.7%)
	Fundoscopy changes	7 (58.3%)
	Other symptoms	8 (66.7%)
Location of pathology		
	Lateral orbital	6 (50%)
	Medial orbital	7 (58.3%)
	Inferior orbital	5 (41.7%)
	Superior orbital	6 (50%)
	Intracranial	1 (8.3%)
	Intraconal	11 (91.7%)
	Intraocular	0
	Attachment to optic nerve	5 (41.7%)
Duration of symptoms at presentation (mo)		14.8
Recurrence		7 (58.3%)
Mean f/u (mo)		28.7
Survival		10 (83.3%)

Abbreviations: EOM, extraocular muscle; f/u, follow-up.

Recurrence

The overall prognosis of orbital paragangliomas is good, but patients with local recurrences have an increased risk of distant metastasis and mortality. Both mortalities that occurred in the population studied were secondary to tumor recurrences. In one patient, the tumor recurred with distant metastasis to the vertebra and the patient passed away as a result of disease. The second patient presented with large neck and orbital masses that were incompletely resected surgically. Eighteen months later, both masses started to enlarge and the patient succumbed to his disease.^{3,27} The recurrence rate in our study was 35.7%. Complete resection in patients with advanced disease or intracranial extension is difficult and can result in significant morbidity. Moreover, due to the locally destructive nature of the tumor, differentiating normal tissue from tumor can be difficult; hence, residual tumor is often left

Table 6 Summary of patients who had a recurrence

Clinical features	Subcategory	Totals
No. of patients with recurrence		10 (35.7%)
Age (y)		34.7
Sex (M:F)		6:4
Clinical features		
	Proptosis	8 (80%)
	EOM restriction	7 (70%)
	Visual acuity changes	7 (70%)
	Eye pain	3 (30%)
	Fundoscopy changes	4 (40%)
	Other symptoms	6 (60%)
Location of pathology		
	Lateral orbital	6 (60%)
	Medial orbital	4 (40%)
	Inferior orbital	4 (40%)
	Superior orbital	5 (50%)
	Intracranial	1 (10%)
	Intraconal	8 (80%)
	Intraocular	0
	Attachment to optic nerve	4 (40%)
Mean duration of symptoms at presentation (mo)		14.9
Loss of vision		7 (70%)
Surgical complication		7 (70%)
Mean f/u (mo)		29.6
Survival		8 (80%)

Abbreviations: EOM, extraocular muscle; f/u, follow-up.

behind.³⁰ Patients with tumor recurrences have a higher rate of tumor attachment to the optic nerve (40 vs. 28.6%) and the one patient in our study who had intracranial extension of their tumor also developed a recurrence. In addition, patients who developed recurrences had an increased postoperative complication rate (70 vs. 60.7%), including loss of vision (70 vs. 42.9%). This illustrates the need for close follow-up for these patients using serial CT scans, especially in patients with residual tumor postoperatively.

Limitations

As a systematic review, one of the limitations of our study is a selection bias, as only studies that met inclusion criteria were included in the final analysis. In addition, as the topic studied is rare, all of the data come from case reports which can include selection and temporal biases. There have been no

randomized controlled trials conducted for this disease. This study could be enhanced if more information was available on tumor volume. Finally, some reports contained limited follow-up time which limits the ability to accurately measure recurrence rates.

Conclusion

This study is the largest systematic review on orbital paragangliomas in the English literature. Orbital paragangliomas are benign tumors in most cases. The most common presenting symptom is proptosis. The primary treatment modality is surgery; usually only tumor excision is required with exenteration reserved for more advanced cases. There is a 35.7% recurrence rate and patients with recurrence are at increased risk of dying from their disease. Patients who had tumor attached to the optic nerve had an increased risk for recurrence and complications, including visual loss, which suggests that these patients may require more frequent and longer clinical follow-up.

Financial Disclosure

None.

Conflict of Interest

None.

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