Malignant Head and Neck Paragangliomas: The M.D. Anderson Experience

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ABSTRACT

Paragangliomas (PGs) - rare tumors of neuroendocrine origin
- Malignant PGs - paraganglioma cells in lymph nodes or in distant metastatic disease
- Small percentage develop metastasizing disease, and represent only 4% to 15% of all head and neck PGs
- Rates of malignancy have been reported as 2% for jugulo-tympanic tumors, 18% for cervical body tumors
- Regional disease, radiation after primary surgical treatment offers best prognosis
- For distant metastases, mortality and lung and liver, are treated with chemotherapy
- Most studies PG with distant disease are based on thoracic region
- Abdominal and thoracic PG originizes in sympathetic autonomic ganglia
- Head and neck paragangliomas originate in parasympathetic ganglia
- To define the outcomes and optimal treatment strategies of malignant head and neck PG, we reviewed a single institution's 35 year experience with these lesions

METHODS AND MATERIALS

All patients with metastatic paragangliomas of the head and neck treated at the University of Texas M.D. Anderson Cancer Center (MDACC) from 1970 to 2005 were reviewed after IRB approval was obtained. Pathology, radiologic imaging, operative reports, and all clinical notes were reviewed. Patients referred to our institution for recurrent or progressive disease were included. All patients had primary surgery at outside hospitals and were referred to MDACC for recurrence or progression. All patients had treatment of their primary tumor at an outside facility. Locoregional and distant treatment approach were revisited. Progression, patient survival, and complication rates were evaluated. P-value was determined by Fisher's exact test. Results: All patients with age over 40 years (N = 19) had distant progression of disease. Of the patients 40 years or less (N = 20), 2 patients had no progression of disease and 1 patient had regression of disease. Patients without progression of disease had better prognosis and all were alive at last follow-up. Although male patients had a higher rate of disease progression with treatment (80%) than female (70%), there was not a significant difference. Conclusions: Malignant paragangliomas accounted for 18% of primary head and neck paragangliomas. Moderate to severe progression was seen among patients less than 40 years-old. With an overall survival of 86% and 53% at 5 and 10 years respectively, patients may benefit from aggressive treatment strategies.

INTRODUCTION

Objectives: To determine prognosis and survival of patients with malignant paragangliomas of the head and neck. Malignant paragangliomas are rare tumors of neuroendocrine origin. A retrospective case review analysis was conducted from 113 cases of head and neck paragangliomas at M.D. Anderson from 1970 to 2005. Twenty patients were lost to follow-up in the regional or distant metastatic disease. One patient was excluded from the study due to 36% follow-up. The mean follow-up was 12 years. All 79 patients had primary surgery at their primary tumor at an outside facility. Locoregional and distant treatment approach were revisited. Progression, patient survival, and complication rates were evaluated. P-value was determined by Fisher's exact test.

RESULTS

RESULTS

Epidemiologic Data
-113 patients had cervical paragangliomas, 23 (18%) had regional or distant metastases
-All tumors were 35 year experience with these lesions

CONCLUSIONS

- Therapy for metastatic head and neck paragangliomas remains controversial
- No treatment demonstrated therapeutic superiority
- Aggressive treatment with radiation therapy and systemic chemotherapy did not achieve disease control in most patients
- Further investigations will be needed to capture the indolent and complex course these tumors typically experience

REFERENCES


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