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Ocular Metastases of Neuroendocrine Tumors

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BACKGROUND: Advances in somatostatin receptor scintigraphy with 68G-dotatate PET scanning can identify metastases in unusual locations, including ocular metastases (OMs) and provide information on response to therapy. Here, we report our experience with OM patients at a neuroendocrine referral center.

METHODS: We identified 6 NET patients with OM seen from April 2012 to April 2019. Charts were reviewed for demographics, clinical syndromes, symptoms, NET history, and follow-up after receipt of Institutional Review Board approval.

RESULTS: Table 1 describes the patients. Median time from initial NET diagnosis to OM was 13.8 years; one had OM diagnosed 15 months before identification of the primary lung tumor. 5 were symptomatic with strabismus +/- visual loss. The asymptomatic patient was incidentally found to have OM on MRI after routine ophthalmologic exam suggested OM. None were identified with gallium scan alone. Five (83%) had other metastases. 4 (67%) received systemic chemotherapy for NET, 5 (83%) received somatostatin analogue treatment. Two patients (33%) underwent PRRT. One (pancreatic primary) received everolimus / CAP-TEM, TACE for liver metastases, EBRT to bone metastases and 2 cycles PRRT after distal pancreatectomy / splenectomy. The second (lung primary) received carboplatin / etoposide, EBRT of lung and bone, and 3 cycles PRRT. Both underwent EBRT to OMs, with significant improvement despite systemic progression. Of the other 4, 1 had OM surgically resected, without recurrence on MRI and controlled systemic disease. Two had EBRT to OM, with one being definitively treated of both systemic and OM disease, and the other with worsening of disease, including OM. The last was asymptomatic, imaging showed stable lesion despite progressing systemic disease (patient expired).



CONCLUSION: This is the first series to describe NET OM in the era of PRRT. OM is usually diagnosed after identification of the primary tumor in the setting of widespread disease, requiring systemic and metastatic directed therapies.

Table 1: Characteristics of patients with ocular metastases

Age at neuroendocrine tumor diagnosis, years, median (IQR)	66.9 (63.9, 68.8)
Age at ocular metastasis diagnosis, years, median (IQR)	68.1 (65.1, 70.6)
Male	3 (50%)
Primary site	
Lung	3 (50%)
Pancreas	1 (17%)
Small bowel	1 (17%)
Small bowel vs pancreas, unknown	1 (17%)
Grade	
1	1 (17%)
2	4 (67%)
3	1 (17%)
Syndrome Related	
None	1 (17%)
Carcinoid	4 (67%)
Insulinoma	1 (17%)
Orbital metastasis related symptoms	5 (83%)*
Metastases to other locations	5 (83%)
Liver	
3 (50%)	
Bone	3 (50%)
Soft tissue	3 (50%)
Systemic chemotherapy	4 (67%)
Somatostatin analogs	5 (83%)

Table 1 (continued)

External beam radiation therapy	4 (67%)
Peptide Receptor Radionuclide Therapy	2 (33%)
Ocular specific treatment	
External beam radiation therapy	5 (83%)
Resection	1 (17%)
Outcome of ocular metastasis	
Treated successfully	2 (33%)
Significant improvement	3 (50%)
Stable	1 (17%)
Surviving at last follow-up	4 (67%)

*Patient without symptoms had routine ophthalmologic exam concerning for metastasis, prompting imaging