

Neuroendocrine metastasis to the orbit – a large multi-centre retrospective case series

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Introduction: Neuroendocrine neoplasms most frequently with primary sites found within the gastrointestinal tract and bronchial tree can metastasize to the orbit. These are very rare, with less than fifty published cases, resulting in potential diagnostic challenges. We present a large, multi-centre retrospective case series.

Method: Multicentre retrospective case note review. Histopathological reports and clinical letters from 1998 to 2016 carried out at Moorfields Eye Hospital, the UCL Institute of Ophthalmology and Addenbrooke's Hospital were searched for using the terms "carcinoid" and "neuroendocrine". Case notes were reviewed and details summarized.

Results: 11 cases were identified (2 female, mean age 68.9 years). 6 cases involved metastases to one or more of the recti muscles, 1 involved infiltration of the lacrimal gland and the remainder were biopsied from other orbital sites. 2 cases were initially diagnosed as having thyroid eye disease (TED), with the diagnosis of carcinoid being made following orbital biopsy and staging CT imaging demonstrating the primary lesions (small bowel). 2 cases were diagnosed based on radiological appearance and neuroendocrine specific imaging, with known history of histologically confirmed primary ileocaecal neuroendocrine malignancy, and therefore not biopsied.

Discussion: Although rare, it is important to consider carcinoid metastases as a possible differential diagnosis for an orbital mass. It should also be considered in an atypical case of TED as the clinical signs of "carcinoid syndrome" can be similar to hyperthyroidism, and the clinical and radiological appearance may cause diagnostic confusion.

Neuroendocrine orbital metastasis may occur to various orbital sites, presenting in a multitude of ways, from an asymptomatic incidental radiological finding, to proptosis, diplopia, visual disturbance and systemic features. Not all cases require biopsy if there is a known primary neuroendocrine diagnosis and the patient is asymptomatic. Neuroendocrine specific imaging can also aid diagnosis. Treatment is often systemic, but can involve localised excision or debulking biopsy in necessary cases.