

Case Study

ORBITAL PROPTOSIS- A RARE PRESENTATION OF THYROID CARCINOMA: CASE REPORT AND REVIEW

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ABSTRACT

A 70 years female presented with right sided orbital proptosis since one and half years with past history of thyroidectomy. On examination thyroid swelling was present with right orbital proptosis. Routine blood investigations were normal except Thyroglobulin levels, 22000IU. Histopathological examination (HPE) of the biopsy from the protruding mass from the right orbit showed follicular acini lined with cuboidal cells. CECT (Contrast enhanced computerized tomography) of orbit showed mass in the superior part of retro orbital region eroding the superior wall and extending into right parasellar. A total thyroidectomy was performed and HPE of specimen was suggestive of thyroid papillary carcinoma. For the orbital metastasis patient underwent external beam radiotherapy to orbit with 40 Gray in 20 fractions. During her follow up period she was found to have secondary in the left femur metaphysis. Patient refused surgical excision of bone secondary. Patient is on regular follow up with supportive treatment.

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INTRODUCTION

Thyroid carcinoma is the most common endocrine malignancy with increasing incidence over the past decade world wide(1). Papillary carcinoma thyroid is the most common form of thyroid malignancy with distant metastasis being uncommon, commonly involving lung and bone, rarely involves the orbit (2). Orbital metastasis are infrequent etiology of adult proptosis. Here we present a case of papillary thyroid carcinoma who presented with unilateral orbital proptosis due to metastasis.

CASE REPORT

A 70 years female presented with right sided orbital proptosis since one and half years (Fig 1) with diminution of vision and swelling in front of neck since 1month. A history of thyroid surgery 40 years back (details were not available) was present. Since then she was on suppressive dose of thyroid hormone and calcium supplementation.

On examination a solitary swelling in front of the neck around 3x4cm, moving with deglutition, consistent with thyroid swelling and right orbital proptosis were present. Routine blood investigations were within normal limits and thyroid profile was in normal range. Thyroglobulin levels were elevated 22000IU. Histopathological examination of the biopsy from the protruding mass from the right orbit showed follicular acini lined with cuboidal cells with tumor cells staining positive for Thyroid Transcription Factor-1 (TTF-1). CECT of orbit showed mass in the superior part of retro orbital region eroding the superior wall and extending into right parasellar region (Fig3 Fig4). A total thyroidectomy was performed. Cut surface of the thyroidectomy specimen showed multiple colloid filled nodules of varying size with areas of haemorrhage with microscopic features of nodular hyperplasia with cystic changes and haemorrhage with a nodule showing sheets of oncocytic cells and small foci of micropapillary carcinoma amidst nodular thyroid tissue one with typical papillary features: diagnostic of papillary carcinoma thyroid with intrathyroidal spread and nodular hyperplasia thyroid with cystic changes and haemorrhage. For the orbital metastasis patient underwent external beam radiotherapy (EBRT) to orbit with 40 Gray in 20 fractions. Following thyroidectomy and EBRT, serum thyroglobulin level declined.

During her follow up period she started developing pain and limping of the left lower limb and her serum thyroglobulin level started to raise. X-ray revealed a solitary lytic lesion in the metaphysis of the femur suggestive of secondary. Patient refused surgical excision of bone secondary. Patient is on regular follow up with supportive treatment.



Figure 1:Pre operative picture showing right eye proptosis and thyroid swelling.



Figure 2: Right eye after external beam radiotherapy

lung (6, 7). Other less frequent primaries found to do so are in the kidneys, testicles, prostate, and bowel (3, 5-7).

Metastatic thyroid carcinoma rarely involves the orbit. They constitute 5% to 6.5% of the total orbital neoplasms (8). Rarely thyroid carcinoma has to be considered as a potential primary tumor in a patient with an orbital metastasis.

In the orbit there are no lymph nodes and no lymphatic vessels have been demonstrated. Probably the main lymph drainage from the orbit accompanies the veins through the inferior orbital fissure to the internal maxillary nodes; thence to the superior deep cervical nodes. A study using radioisotope thyroidolymphography and orbitolymphography suggested a probable lymphatic channel connection between the thyroid gland and the orbit (9). Such thyroido-orbital lymphatic channel anastomoses may be the route for orbital deposition of thyroglobulin-antithyroglobulin immune complexes with Graves's ophthalmopathy (10). The apparent predisposition of thyroid carcinoma to orbital rather than ocular spread could be compatible with proposed thyroido-orbital pathway.

Orbital metastases are an infrequent aetiology of adult proptosis; approximately 3–7% of orbital biopsies have demonstrated a metastatic tumour, and this diagnosis is often unexpected (11). Metastatic orbital deposits are less common than uveal metastases; in several series the relative incidence was approximately one to eight (12,13). In contrast to other malignancies metastasising to eye, with thyroid carcinoma the pattern is reversed: thyroid carcinoma metastases to the orbit are commoner than to the globe (14). In both sites, a metastatic tumor may present with ophthalmic symptoms before the discovery of the primary neoplasm; this more common with lung, gastrointestinal, thyroid, and renal carcinomas (12).

Most commonly, patients present with diplopia (48%); less frequently proptosis (26%) and decreased vision (16%) less commonly pain and ptosis (15). Diplopia was the most common presenting symptom in patients with metastatic tumours, in contrast with most other orbital neoplasms, where either proptosis or visual loss was more frequent (11). Unfortunately, this symptom array is not diagnostic; in an older patient with mild proptosis and diplopia and no systemic thyroid signs or symptoms, the possibility of a metastatic orbital tumour should be considered in the differential diagnosis.

Most commonly the muscle and contiguous orbital structures are involved with metastatic foci; involvement of the muscle or muscles alone is less common. It is estimated that approximately 5% of orbital metastases involve only extraocular muscles (17). In as many as 25% of choroidal metastases, multiple or bilateral tumours were present; bilaterality is much less frequent in orbital metastases.

Even with modern non-invasive diagnostic tests there is a significant false negative diagnostic rate for the detection of a metastatic aetiology of these orbital lesions. MR (magnetic resonance) data are not superior to CT to establish the diagnosis of metastases. Several patterns are thought to be very typical for orbital metastases, especially with a history of a systemic malignancy. These included intramuscular focal masses, bone destruction plus contiguous mass, and diffuse intraconal lesions (15).

Therapy is based on establishing a correct diagnosis, the systemic status of the patient, and whether optic nerve compression is present. In patients with suddenly decreased vision due to a mass compressing the optic nerve, surgical debulking of the tumour is the optimum treatment and it is possible to restore vision in such patients. In a few patients with focal tumours that produce symptoms from a mass effect, surgical extirpation of the lesion is locally effective (15).

In patients with a solitary orbital metastasis that is either diffuse or involved an important structure, such as the muscle, globe, or nerve, almost 80% of patients will obtain symptomatic relief with a total dose of 30 to 40 Gy of photon radiation. It is imperative to obtain a brain MR scan with gadolinium before irradiating a metastatic orbital tumour (18). Not infrequently, patients will have silent brain lesions when they present with orbital disease. In patients who have both widespread metastases and chemotherapeutically sensitive tumors, chemotherapy is the treatment of choice.

As other series have shown, prognosis is usually dismal in these patients; however, a few have had long term remissions and are alive 10 years after the orbital disease was diagnosed (12,14).

The median survival is a little over 1 year, and only 27% had 2 year survival. Patients with breast cancer have a slightly longer survival than patients with other malignancies (14,19).

Survival differences with choroidal versus orbital metastases, and different histological types of orbital metastases that have been described must be interpreted cautiously. In some tumors, such as renal carcinoma, cases have been reported with long term survival after removal of a solitary orbital metastasis.

CONCLUSION

Orbital metastasis as a primary manifestation of papillary carcinoma thyroid is very rare event. Unilateral proptosis in elderly age group should always be investigated as a distant metastasis.

COMPETING INTERESTS

No competing interests exist.

AUTHORS' CONTRIBUTION

Author 1 did the first draft of the manuscript and literature searches.

Author 2 and Author 3 involved in patient workup and treatment of the patient.

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