## Malignant transformation of ectopic thyroid tissue in the orbit: A rare case report

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#### **ABSTRACT**

Ectopic thyroid tissue refers to the presence of thyroid tissue in locations other than normal anterior neck region. It is the most frequent type of thyroid dysgenesis. Lingual thyroid is the most common type accounting for 90% of the cases. The presence of thyroid tissue in orbit is extremely rare along with malignant transformation and the presence of normal thyroid gland. We report a very rare case of follicular carcinoma arising in the ectopic thyroid tissue in the left orbit along with normal thyroid gland. To the best of our knowledge, it is the first ever reported case of this type.

Key words: Ectopic thyroid tissue, follicular carcinoma, normal thyroid gland

#### **INTRODUCTION**

Ectopic thyroid tissue refers to the presence of thyroid tissue in locations other than anterior neck region between the second and fourth tracheal cartilage. <sup>[1]</sup> It is the most frequent type of thyroid dysgenesis accounting for 48–61% of cases. <sup>[2]</sup> The prevalence of this condition is between 1/100,000 and 300,000. <sup>[3]</sup> Hickman reported the first ever case of ectopic thyroid tissue present at the base of tongue pressing down the epiglottis on the larynx causing death by suffocation sixteen hours after birth. <sup>[4]</sup> We report the case of a 60-year-old male with follicular carcinoma arising in ectopic thyroid tissue in the left orbit with normal eutopic thyroid.

#### **CASE REPORT**

A 60-year-old male presented with complaints of pain, diminution of vision, and swelling in the left eye. On

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examination, there was proptosis and diminished vision. He was subjected to a battery of investigations. In the present case contrast, magnetic resonance imaging showed approximately 46 (AP) × 33 (CC) × 42 (TR) mm sized well-defined lobulated mass in extra-axial space in superior quadrant of left orbit. The mass was causing the destruction of the left lesser wing of sphenoid extending to middle cranial fossa and indenting anterior temporal lobe cortex. Superiorly, there was the destruction of orbital roof and indenting under the surface of the frontal lobe. Laterally, there was the destruction of zygomatic bone and extending to left frontal scalp. The mass was causing displacement of orbital contents inferiorly with proptosis of the left eyeball [Figures 1-4]. Complimentary ultrasonography screening of eutopic thyroid gland showed normal study. The T3, T4, and thyroid-stimulating hormone levels were normal. The patient was operated on, and the tissue from the left orbit was subjected to histopathological examination. Hematoxylin and Eosin-stained sections were examined.

Sections studied from the retrobulbar tumor showed thyroid tissue with solid, microfollicular and macrofollicular

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architectural pattern. The tumor cells displayed a large highly pleomorphic nuclei, with the presence of nucleoli and scant to moderate amount of cytoplasm. At places, the tumor cells were invading the stroma as well as the capsule [Figures 5 and 6]. Immunohistochemistry was done on tumor sections which showed positivity

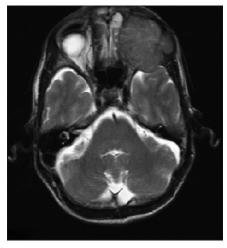
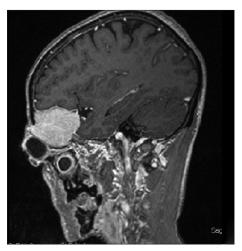


Figure 1: Magnetic resonance imaging scan of skull showing well-defined lobulated mass in extra-axial space in superior quadrant of left orbit (axial T2)



Figure 2: Magnetic resonance imaging scan of skull showing well-defined lobulated mass in extra-axial space in superior quadrant of left orbit (post Gd coronal)



**Figure 3:** Magnetic resonance imaging scan of skull showing well-defined lobulated mass in extra-axial space in superior quadrant of left orbit (Sag T1)

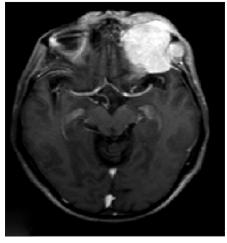
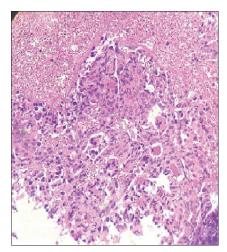
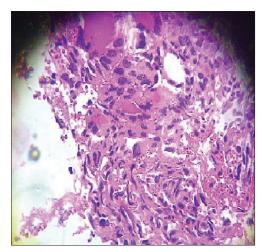


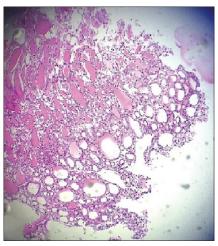
Figure 4: Postcontrast magnetic resonance imaging scan of skull showing well defined lobulated mass in extra-axial space in superior quadrant of left orbit (axial)



**Figure 5:** Photomicrograph showing pleomorphic malignant follicular epithelial cells arranged in macrofollicles and microfollicles (H and E, ×100)



**Figure 6:** Photomicrograph showing pleomorphic malignant follicular epithelial cells with moderate cytoplasm and prominent nucleoli (H and E, ×400)

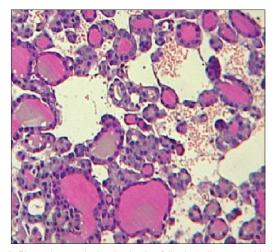


**Figure 7:** Photomicrograph showing benign follicular cells arranged in follicles with abundant colloid (H and E, ×100)

for thyroglobulin and thyroid transcription factor-1. Nontimorous areas revealed normal thyroid follicles lined by uniform follicular cells with round to oval nuclei, scant cytoplasm, and abundant colloid [Figures 7 and 8].

#### **DISCUSSION**

Ectopic thyroid tissue refers to the presence of thyroid tissue in locations other than the normal location of thyroid, i.e., the anterior neck region between the second and fourth tracheal cartilage.<sup>[1]</sup> Mutations in the TITF-1, Foxe1, and PAX-8 may be involved in the abnormal migration of the thyroid gland. [2] Lingual thyroid is the most common type of ectopic thyroid accounting for 90% of all the cases. Sublingual thyroid tissue is less frequently encountered.[1] The other locations in the head and neck region where ectopic thyroid tissue can be found are the iris of the eye, pituitary gland, carotid bifurcation, tonsils, submandibular gland, trachea, and the lateral cervical region. Primary malignant transformation in ectopic thyroid tissue is very rare, and diagnosis is based on the histopathological criteria similar to eutopic thyroid glands. Papillary carcinoma, follicular carcinoma, follicular variant of papillary carcinoma, mixed follicular and papillary variants, Hurthle cell carcinoma, and medullary carcinoma have been reported in ectopic thyroid tissues.<sup>[5,6]</sup> The majority of the tumors that have been described in the ectopic thyroid are the follicular carcinomas whereas papillary carcinomas comprise only 23%, which contrasts to the predominance of papillary carcinomas in the eutopic thyroid gland.[6]



**Figure 8:** Photomicrograph showing benign follicular cells arranged in follicles with abundant colloid (H and E, ×400)

#### CONCLUSION

We can conclude that the presence of thyroid tissue in orbit is extremely rare. Malignant transformation of the ectopic orbital thyroid with normal thyroid gland is rarest of rare, and to our belief, this is the first reported case.

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#### **Conflicts of interest**

There are no conflicts of interest.

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