



Clinical Spectrum and Surgical outcome of Follicular Variant of Papillary Carcinoma of Lingual Thyroid with Nodal Metastasis in Neck and Absent Orthotopic Thyroid Gland

(Case Report)

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Abstract

Lingual thyroid is relatively uncommon developmental defect of thyroid gland embryogenesis. We report a case of 48 year-old female patient who presented to us with swellings in both side of neck since 2 years. She was diagnosed follicular variant of papillary carcinoma of lingual thyroid with nodal metastasis and absent orthotopic thyroid tissue. Till date less than 50 cases of lingual thyroid carcinoma have been described in the international literature. To the best of our knowledge, less than 3 cases of this variant have been reported. Treatment was managed with surgical excision of all thyroid tissue followed by radioactive iodine ablation and levothyroxine therapy. This case report provides current understanding about the clinical spectrum of this rare condition, also referring to optimal diagnostic approach including role of MRI and management strategies.

Key-words: Follicular variant, Papillary Carcinoma, Lingual Thyroid, Neck Metastasis

Introduction

Lingual thyroid results from the failure of thyroid primordium to migrate from its embryonic site of origin, the foramen cecum to its normal anatomic position in the neck. Its prevalence is about 1 per

100 000–300 000 people, rising to 1 per 4000–8000 in patients with thyroid disease^[1]. Hickman first described a case of lingual thyroid in 1869. Symptomatic lingual thyroid tissue is unusual with approximately 400 previously

reported cases. Carcinoma of the lingual thyroid is a very rare clinical entity with an estimated incidence of 1%. The case of Lingual thyroid carcinoma was first reported by Gunn and by Rutgers, independently in 1910 [2]. Follicular carcinoma of the lingual thyroid is the commonest histopathological subtype [4]. There is currently no explanation for the apparent follicular carcinoma predominance in cancers of the lingual thyroid which contrasts with carcinomas of orthotopic and ectopic thyroid tissue where papillary carcinoma dominates [4]. This case report provides current understanding about the clinical spectrum of this rare condition, also referring to optimal diagnostic approach including role of MRI and management strategies.

Case History

A 48 year-old female patient presented to us with swellings in both side of neck since 2 years [Figure 1: MRI images of patient with multiple lesions in favour of encephalitis, 1a: Axial image on T1 weighted sequence, 1b: Axial image on T2 weighted sequence, 1c&1d: Coronal and axial image FLAIR sequence, 1e: Axial image on T1 W post contrast sequence]. The swellings gradually increased in size causing dysphagia. Patient had history of dysphonia, cough, mild fever, weakness and loss of appetite since 3 months. There was no history of pain from the swelling or any symptoms of airway obstruction. No significant family history was present.

On examination, swelling was firm in consistency extending to midline of neck. Skin was dry. Blood pressure was 112/80mm of Hg. Thyroid function test suggested primary hypothyroidism with elevated TSH levels [100 mIU/L (Normal level 0.4–4.0 mIU/L)], low T4 levels [14.6 nmol/L (57.9–161)], and low T3 levels [659 nmol/L (1.25–2.74)]. FNAC from neck swelling was done which showed clusters of follicles and isolated as well as clump of epithelial cells showing hyperchromatic nucleus, scanty cytoplasm, prominent nucleolus. Numerous eosinophilic hyaline globules were also seen at places. All

features were suggestive of secondaries from thyroid carcinoma [Figure 2: MRI imaging after 3 months, showing multiple enhancing lesions arising suspicion of multicentric gliomas. 2a: Axial image on T2 weighted sequence, 2b: Coronal image on T1 W post contrast sequence, 2c: Axial image on T1 W post contrast sequence]. She underwent MRI which showed a heterogeneous mass at base of tongue which appeared iso to hypointense on T1W sequence [Figure 3: After 1 month of previous MRI, follow up MRI imaging suggesting of multicentric gliomas

3a: Axial image on T1 weighted sequence] and predominantly hyperintense on T2W sequence [Figure 3b Axial image on T2 weighted sequence&3c: Axial image on FLAIR sequence]. Two masses appearing hypointense on T1W sequence [Figure 4: : Histopathological section showing Glioblastoma Multiforme, WHO Grade IV (H&E, 100X)] and predominantly hyper intense on T2W sequence [Figure 5: Postoperative MR imaging showing post operative changes in the form of craniotomy defect (red arrow), hemorrhage (white arrow), residual lesion in right frontal lobe (green arrow) and lesion with internal necrosis in left frontal lobe (blue arrow) which increased in size compared to preoperative MRI. 5a: Axial image on T1 weighted sequence, 5b: Axial image on T2 weighted sequence, 5c: Axial image on FLAIR sequence, 5d: Axial image on T1 W post contrast sequence] were seen in both side of neck which were displacing adjacent blood vessels and soft tissue structures with maintained fat planes. Thyroid gland was not visualised at its normal anatomical position. Forceps biopsy of lingual mass showed exclusive numerous true papillae comprising of variable sizes of follicles. The follicular epithelial cells were having high N:C ratio, scanty cytoplasm with optically clear nuclei and nuclear grooves. At some places invasive growth pattern with fibrous trabeculations and strongly eosinophilic colloid material were also seen. These findings were suggestive of follicular

variant of papillary thyroid carcinoma. [Figure 6]. Further confirmation of lingual mass was done by immunohistochemical examination which showed positive immunoreactivity for thyroglobulin and TTF-1. Hence, final diagnosis was confirmed as follicular variant of papillary carcinoma of lingual thyroid with nodal metastasis and absent orthotopic thyroid gland.



Fig 01

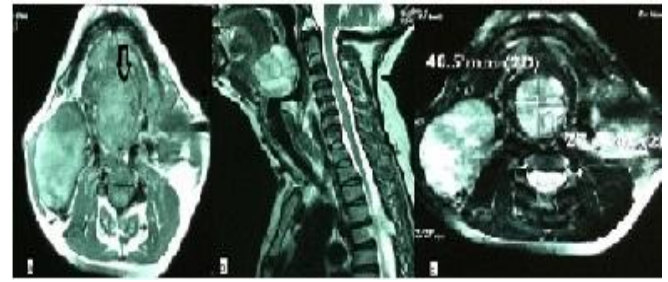


Fig 04

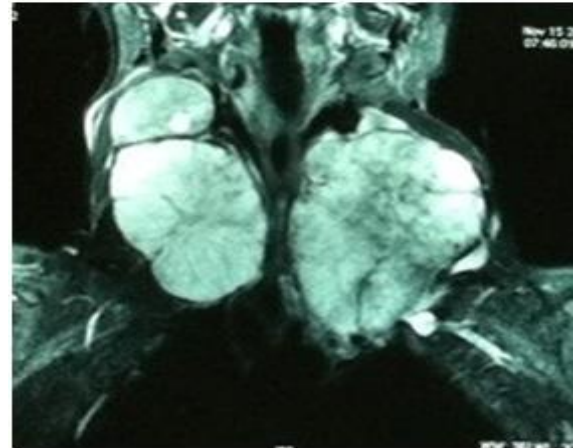


Fig 05

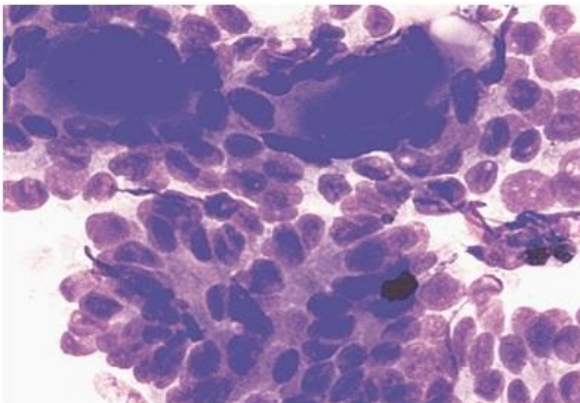


Fig 02

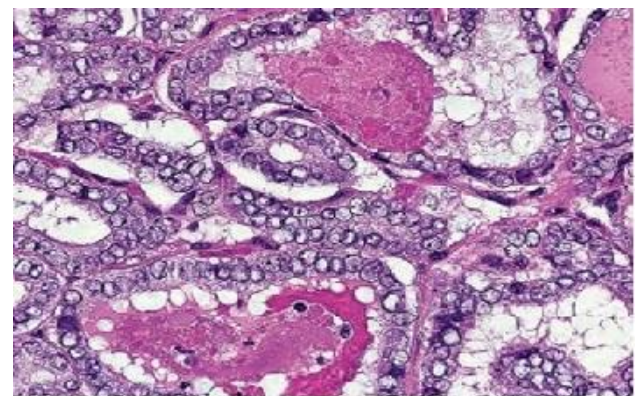


Fig 06



Fig 03

Treatment

There is no consensus about the optimal therapeutic strategy, perhaps due to the rarity of this clinical entity. In this patient, the lingual lesion was excised surgically with a margin of surrounding normal tissue through a lateral pharyngotomy approach with bilateral neck dissection. A tracheotomy was performed to ensure a safe airway. The patient recovered uneventfully from the surgery. Thereafter, patient was given radioactive iodine therapy and levothyroxine suppression, with subsequent

follow-up after 6 months with serum thyroid-stimulating hormone and thyroglobulin assay , TSH level were within normal limits (3.4 mIU/L) and blood thyroglobulin was undetectable.

Discussion

The origin of a lingual thyroid is the result of a failure of the thyroid primordium to descend into the neck. As a consequence, a focus of thyroid tissue remains at the foramen cecum resulting in lingual thyroid. On the other hand, an over descent of the thyroid primordium can result in the formation of ectopic thyroid tissue in the mediastinal area or even within the cardiac endothelium. The majority of complaints in lingual thyroids arise during puberty, pregnancy or menses with an average age of 40 years. However, it may appear at any time from birth to old age . Symptoms are largely because of mass effect and include dysphonia, dyspnoea, haemoptysis, or a foreign body sensation.

Studies of Montgomery's 30 and Buckman's 5 reports reveals that only 30 per cent of reported cases were thought to have thyroid present in the normal position. Therefore, between two thirds and three quarters of patients with symptomatic lingual thyroid will have no other functioning thyroid tissue.

Out of these approximately 70% of patients will be hypothyroid as was this patient . However, they may also be euthyroid, even when no orthotopic thyroid exists [8].

Our review of the literature found fewer than 50 cases of a malignant change in an ectopic lingual thyroid, Follicular carcinoma of the lingual thyroid is the commonest histopathological subtype [4]. Other histopathological types described in ectopic thyroid includes mixed follicular, and papillary Hurthle cell and medullary carcinomas [5,6]. There is a slight female predominance similar to that found in carcinoma of normally placed thyroid tissue in which an approximate 2:1 female-to-male ratio exists.

The carcinoma of ectopic thyroid tissue do not differ in cell types from those arising in orthotopic

thyroid gland [3]. This case had follicular variant of Papillary carcinoma.

Various subtypes of Papillary thyroid carcinoma have been identified. These include follicular, encapsulated, diffuse sclerosing, columnar cell and tall cell types.

Chem and Rosai described the entity called 'follicular variant of papillary carcinoma' in 1977. Papillary carcinomas having an exclusive or almost exclusive follicular pattern are designated as a follicular variant of papillary carcinoma. The biologic behaviour of this variant is analogous to that of conventional papillary carcinoma. The metastases may have a mixed papillary and follicular formation. The immunophenotypic profile of follicular variant of papillary carcinoma is similar to that of classical PTC and is different from follicular carcinoma. Follicular variant also has a similar tendency for lymph node metastasis as that of classical papillary thyroid carcinoma.

It difficult to suggest an accurate treatment strategy or a precise prognosis of this variant because of rarity of this type of tumour in lingual thyroid, along with the lack of information about its natural history and final outcome in most of the treated patients. The same principles that is applied for treatment of an orthotopic thyroid gland papillary thyroid carcinoma is also valid for lingual thyroid papillary tumours including the follicular variant.

According to the current management guidelines for patients with differentiated thyroid cancer issued by the American Thyroid Association , treatment options includes surgery, radiotherapy, radioactive iodine therapy, and chemotherapy [7]. Several surgical approaches for lingual thyroid have been described, such as the transoral route and the transhyoid, suprahyoid, or lateral pharyngotomy approach. The transoral route is usually preferred for small lesions since it does not affect deeper structures; thus, complications, such as lingual nerve injury and deep cervical infections, are avoided. Pharyngotomy with or without preoperative tracheotomy is chosen for larger masses providing better control of bleeding

[5,9,10]. Adequate surgical treatment followed by postoperative radioactive iodine ablation can offer very promising results for ectopic papillary thyroid carcinoma. Treatment outcomes of papillary thyroid carcinoma are very favourable and have an excellent prognosis with 10-year survival rates of more than 90% .

Conclusion

Our presently reported case shows that there is always a chance to encounter exceedingly rare disease. This case report provides understanding of clinical spectrum, optimal diagnostic approach including role of MRI and outcome of surgical management with lateral pharyngotomy approach and bilateral neck dissection in this entity . We believe that this case report could be important for further observation of similar cases in the future.

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