Papillary carcinoma in a supra hyoid thyroglossal duct cyst – an unusual presentation

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Abstract Carcinomatous transformation of ectopic thyroid elements within the thyroglossal tract is a rare entity. We report a case of primary papillary carcinoma of thyroid presenting within the thyroglossal duct cyst in a 23 year old gentleman. The patient presented to us, as a case of supra-hyoid thyroglossal cyst with sub-lingual involvement and he underwent surgical excision of the entire thyroglossal tract by Sistrunk's approach. The post-operative histopathological diagnosis was of a papillary carcinoma within the thyroglossal duct cyst. Hence, the patient was evaluated for a malignant focus in the thyroid which proved negative. He was counseled regarding the possibility of developing Carcinoma in the thyroid gland and offered two options of further management viz., total thyroidectomy followed by life long thyroid suppression or watchful observation and follow up. As the patient belonged to the low risk group, he opted for the second option. He is presently kept under meticulous follow up and remains asymptomatic till date. We present the pre- and post- operative imaging, intra-operative findings, histo-pathological features and review the present world literature on this rare entity.

Keywords Papillary carcinoma thyroid · Thyroglossal duct cyst · Ectopic thyroid · Sistrunk's surgery · Radio iodine nuclear imaging

Introduction

Embryologically, the thyroglossal tract descends from the foramen caecum at the junction of anterior 2/3rd and posterior 1/3rd of tongue into the soft tissues of the neck upto the thyroid bed, by the seventh week of the intrauterine life. Thyroglossal duct cyst is the most common congenital anomaly that arises from the remnants of the thyroglossal duct and its incidence is around 0.5 to 3% in the general adult population.

Nearly 90% of cases of ectopic thyroid tissue reported world wide, were found in the base of the tongue presenting as Lingual Thyroid and the rest 10% were located in the anterior aspect of the neck superior to the hyoid bone. Recent world literature quotes the incidence of papillary carcinoma arising in a thyroglossal duct cyst to be less than 1% and most of these tumors primarily arose from the ectopic thyroid tissue within the cyst [1]. Brentano was the first to describe this condition in 1911. Since then about 215 cases have been reported in world literature [2]. Papillary carcinoma is the most common type of malignancy reported in ectopic thyroid tissues. The prognosis of papillary carcinoma arising within a thyroglossal cyst remains good and quite similar to that of a primary papillary carcinoma of the thyroid gland having cure rates in excess of 95% [2]. Preoperative evaluation of thyroglossal duct cyst should mandatorily include a thorough head and neck examination and thyroid function tests.

A Radio Iodine (I^{123}) Thyroid scan needs to be done if there is suspicion of an ectopic thyroid gland. It is routinely indicated if a tumour mass is palpable within the thyroglossal cyst, or within the thyroid gland and in the presence of metastatic cervical lymph nodes. An ultrasound examination of the thyroglossal duct and the thyroid gland often aids in the diagnosis of a malignancy by demonstrating an intra-mural nodule with calcification or presence of cervical lymph nodal metastases. The final diagnosis of malignancy is most often post-operative, based upon histopathological examination of the excised thyroglossal duct cyst [2].

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Case report

A 23-year-old gentleman presented with a cystic swelling of six months duration in the upper neck just below the chin. A comprehensive ENT and Head and Neck examination revealed that the cystic swelling was present in midline of the upper neck above the hyoid bone and was also seen to extend deep into the floor of mouth. This swelling moved very well with deglutition, but not much with tongue protrusion.

Thyroid function tests were within normal limits and on Ultrasound of the neck, the patient had a normal thyroid gland with both lobes showing normal homogenous echotexture and no evidence of cervical lymphadenopathy. Radio Imaging with High Resolution Computed Tomography showed a 3.8 cm × 3.6 cm size enhancing multi-loculated cystic lesion in the sub-mental region of the neck, with extension into the floor of mouth (Figs. 1 and 2). Polymorphic Calcifications were seen within the capsule of the cystic lesion. FNAC done twice from the swelling proved inconclusive. Since, this cystic lesion was found in the upper neck, in midline extending into floor of mouth and associated with calcifications, the provisional differential diagnosis included a sublingual dermoid cyst, lymphangioma, plunging ranula, thyroglossal duct cyst or a midline teratoma.

Fig. 1 A Computed tomographic scan picture (Axial Section) showing the 3.8 cm × 3.6 cm cystic lesion lying in the sub-mental region, abutting the floor of mouth and base of tongue

After the completion of all the necessary preoperative investigations, the patient underwent excision of the lesion by a Sistrunk's operation. A huge cystic neck mass measuring 3.8 cm × 3.6 cm, extending superiorly upto base of tongue and floor of mouth and inferiorly until the body of hyoid bone was dissected out. The entire lesion was excised en-bloc with the body of the hyoid bone and sent for histopathological examination.

Histo-pathological assessment revealed a diagnosis of papillary carcinoma of thyroid in a supra-hyoid thyroglossal duct cyst. The tumour tissue was well confined within the thyroglossal cyst and the cyst walls were not invaded by the neoplasm. Microscopically, the lesion showed several papillae and thyroid acini with optically clear nuclei. Several areas of calcification were noted and pathognomonic 'Psammoma' bodies were seen confirming the diagnosis of a papillary carcinoma of thyroid (Fig. 3).

The patient was counseled in detail about the pathological findings and in accordance with the existing world literature, two possibilities for further management were discussed with him. The first option was a total thyroidectomy surgery with post operative Radioactive Iodine (I^{131}) Ablation if necessary, followed by life long thyroid suppression. The second option was watchful observation and follow up with no immediate intervention necessary. As stated in the literature, our patient fell in the low risk group and hence he opted out of undergoing a second surgical procedure. He is presently kept under meticulous follow up and is asymptomatic till date. A post-operative CT scan was performed recently at six months follow up and it showed normal healing with no evidence of the lesion (Fig. 4).

Fig. 2 A Computed tomographic scan picture (Sagittal Section) showing the multi-loculated cystic lesion in relation to the Hyoid bone and mandible

Fig. 3 Histopathological photomicrograph of the thyroglossal duct cyst showing the classical appearances of a papillary carcinoma of thyroid with papillary projections lined by malignant thyroid acinar cells and pathognomonic 'Psammoma Bodies' (shown by the arrow)

Fig. 4 Six months post-operative computed tomographic Scan pictures (Axial and Sagittal Sections) showing no evidence of lesion

Discussion

The incidence of papillary carcinoma arising in the thyroglossal duct cyst is less than 1% and most of these tumors arise primarily from the ectopic thyroid elements present within the thyroglossal tract [3]. Present world literature has revealed that, papillary carcinoma is the most frequent carcinoma found in a thyroglossal duct cyst (80%). Other pathological types include follicular or mixed (papillary and follicular) carcinoma (9%), squamous cell carcinoma (5%), adenocarcinoma (2%), anaplastic carcinoma (1%) and others (3%) [4, 10]. Metastasis from the thyroglossal duct cyst malignancy onto the regional cervical lymph nodes is fairly rare, occurring in about 10% of the cases [8]. The prognosis of papillary carcinoma arising in a thyroglossal cyst is very similar to that of papillary carcinoma of the thyroid gland having cure rates in excess of 95% [2].

Our patient predominantly had a midline, sub-mental and intra-oral swelling which is an unusual clinical presentation for a thyroglossal cyst. Hence, in the pre-operative differential diagnosis a thyroglossal cyst was not the first choice. Radio Imaging confirmed the lesion to be suprahyoid, extending upto the base of tongue and well in congruence with the hyoid bone. Due to the above radiological features it was decided that a Sistrunk's operation would provide us the best access to this lesion in order to facilitate its complete removal.

Once the histopathological diagnosis of a papillary carcinoma was made a post-operative Radio Iodine (I^{123}) scan was arranged in order to track any functional elements outside the thyroid bed, especially to look for remnants at the operated site or any metastases. The Radio Iodine (I^{123}) Thyroid uptake scan showed normal uptake in the thyroid gland and no other regional or remote metastasis.

In most of the cases reviewed in recent literature, the suspicion of a malignancy did not arise before the operation and very similar to our case report, papillary carcinoma in ectopic thyroid tissue was diagnosed only at histopathological examination [3]. There remains a lack of consensus in the literature regarding the ideal management for this condition possibly due to lack of a definite pre-operative diagnosis and rarity of this entity [4]. It is now generally accepted that thyroglossal duct carcinoma arises from thyroid tissue remnants located within the cyst itself. While thyroglossal cysts occur most commonly in patients younger than 20 years of age, thyroglossal duct carcinoma occurs more commonly in older patients with an average age close to 40 years. This entity has slight female preponderance and the female to male ratio reported in one review is 3:2 [5]. The most common clinical presentation reported has been an asymptomatic mass, but symptoms may vary and include a rapidly enlarging firm, hard or fixed midline mass with dysphagia, dysphonia, or the presence of cervical lymphadenopathy [5, 8].

Literature has defined two modalities of management depending on the age factor and the histo-pathological features of thyroglossal duct carcinoma [7, 8]. In young patients less than 40 years, with a small tumour well confined within the thyroglossal tract walls, a wide excision by Sistrunk's procedure alone would suffice without any adjuvant therapy. These patients belong to the low risk group and need to be kept under meticulous follow up and watchful observation. Recurrence rate in this low risk group is negligible. Our patient aged 23 years belonged to this low risk category and the histo-pathological picture confirmed the papillary carcinoma to be well confined within the walls of the cyst with no peripheral invasion and with no evidence of regional or remote metastasis.

In older patients beyond 40 years and in larger tumors with erosion of the cyst walls, the preferred treatment

is a wide excision of the entire thyroglossal duct cyst along with the body of hyoid bone, followed by a total or near total thyroidectomy surgery. These patients need to undergo post-operative Radio Iodine (I^{131}) Ablation for remnants, if any and need to be put on life long suppressive doses of thyroid hormones. The necessity for watchful observation and follow up in this high risk group is the inherent multicentric nature of any residual papillary carcinoma which has the propensity for rapid metastasis [6–8] Irrespective of these age and risk stratifications, a total thyroidectomy should always be recommended if there is a suspicious mass in thyroid gland or if a positive history of radiation to the neck is present [6, 8, 9].

Conclusion

Thyroglossal tract may harbor ectopic thyroid tissue, which may have the potency to eventually proliferate into a papillary carcinoma. Formation of papillary thyroid carcinoma within a thyroglossal duct cyst is a rare and unusual entity. Its clinical behavior and prognosis are usually very similar to that of the primary papillary thyroid cancer. In most cases, the diagnosis is often post-operative by histopathological examination. Two fold management options have been described in literature as per the risk stratification, but a final consensus is yet to be achieved regarding the management protocols, due to the rarity of this entity.

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