HIGH-GRADE MUCOEPIDERMOID CARCINOMA IN THYROGLOSSAL CYST: POST-SURGICAL HISTOLOGICAL SURPRISE AND DILEMMAS

Ravindran Chirukandath1, Nimisha CR2, Babu PJ3, Arya Puthukatt Muraleedharan4, Lekshmi RS5, Santhosh TV6

1Additional Professor, 2Assistant Professor, 3Professor and Head, 4Junior Resident, 5Associate Professor, Department of General Surgery, Government Medical College, Thirssur, Kerala, India, 6Senior Resident, Department of Pathology, Government Medical College, Thirssur, Kerala, India

INTRODUCTION

Thyroglossal cyst is the most common congenital abnormality of thyroid gland development affecting pediatric age group but also in older age group.1 Thyroglossal cyst can have ectopic thyroid tissue varied from 1.5% to 45% of cases.2 Occurrence of malignancy in the cyst has been rarely reported, though rare and till date, 278 cases are reported in the literature3 and papillary carcinoma predominates the common type but squamous cell carcinomas, anaplastic carcinoma, and medullary have been reported rarely.

Mucoepidermoid carcinomas are most commonly seen in salivary glands, however, it is also reported from glandular tissue of lung, lacrimal glands, and thyroid. There are around 42 cases of mucoepidermoid carcinomas from world literature and mostly they are low grade. We are reporting the second reported case of mucoepidermoid carcinoma from thyroglossal cyst. This case highlights the importance of removal of thyroglossal duct cysts at an early stage and aggressive surgical approach in high-grade tumors.

Key words: TG cyst; Mucoepidermoid carcinoma; Sistrunk’s operation; High grade

CASE REPORT

Address for Correspondence:
Dr. Ravindran Chirukandath, Additional Professor, Department of General Surgery, Government Medical College, Thirssur, India.
Mobile: +91-9447040431. E-mail: ravimen@gmail.com
of strap muscles. Cytological investigation revealed it to be a TG cyst malignancy. The treatment plan and histopathological surprise that followed is discussed in detail.

PRESENTATION OF THE CASE

A 67-year-old lady presented to general surgery OPD with complaints of a swelling below chin in the upper neck which she noticed 5 months back. Swelling was painless, moving with deglutition and rapidly progressed in size. There was associated discomfort while swallowing. She was known hypertensive on antihypertensives. Clinical examination revealed that there was a 6 × 8 cm sized oval hard non-tender swelling in submental region in midline extending from 2 cm below symphysis menti to just above the level of hyoid bone, and 3 cm to the midline on either side. It was moving with deglutition and with the protrusion of tongue. Thyroid gland was not enlarged. NO palpable cervical lymphnodes. Oral cavity examination was normal.

Ultrasound neck reported as heterogeneous lesion with necrotic collection in submental region in midline measuring 2.5 × 2 × 2 cm with bilateral sub-centimetric level 3 and 4 lymph nodes.

MRI scan was taken which showed an ill-defined heterogeneously enhancing soft-tissue lesion 39 × 37 × 24 mm in midline in submental pre-hyoid region extending to anterior surface of thyroid cartilage (Figure 1) which appears to be embedded in the left strap muscles. Few intralesional cystic areas noted. Lesion was found to be extending posteriorly in pre epiglottic region. Mild bone marrow edema of hyoid bone and thyroid cartilage noted, findings were suggestive of thyroglossal duct cyst lesion with possible malignant infiltration. Thyroid gland visualized normally with no focal lesions.

Fine-needle cytology showed cells suggestive of papillary carcinoma. The patient was prepared was operated. On exploration there a 4 × 5 cm hard lesion above the Hyoid This lesion was found to be infiltrating right strap muscles and body of hyoid. Thyroid appeared to be normal morphology. We proceeded with Sistrunk operation along with total thyroidectomy and Level VI central nodal dissection [Figure 2].

Histological examination revealed a low-grade mucoepidermoid carcinoma consistent with origin in a thyroglossal duct remnant. It invaded the hyoid bone and adjacent strap muscles [Figure 3].

DISCUSSION

Carcinoma arising in a TGDC is a very rare and accounts for less than 1%. Approximately 280 cases of malignant tumors in thyroglossal cyst are reported up to 2020 and rarely thereafter. Most common histological type is papillary carcinoma (80–95%) followed by mixed papillary-follicular carcinoma (8%) and squamous cell carcinoma (6%). It also has to be noted that other cancers such as follicular, squamous cell carcinoma, and Hurtle cell carcinoma are described rarely in the literature. Very rarely there are

Figure 1: MRI shows an ill-defined heterogeneously enhancing soft-tissue lesion 39 × 37 × 24 mm in midline in submental pre-hyoid region extending to anterior surface of thyroid cartilage which appears to be embedded in the left strap muscles.
reported cases of anaplastic carcinoma and one reported case of mucoepidermoid carcinoma. It is very important to emphasize that genetic positivity of TITF1, TITF2, and PAX8 and the gene for the thyroid-stimulating hormone receptor.

The knowledge about the thyroid mucoepidermoid carcinoma is still evolving and it is shown that these low-grade tumors are shown to be positive for the t(11;19)(q21;p13) translocation\(^7\)\(^\rightarrow\)\(^10\) but the lack of this translocation in high-grade tumors points to the fact that these tumors are biologically and molecularly different entity. The histogenesis of thyroid or thyroglossal duct MEC is explained by two theories that it develops from solid cell nests (SCNs) or thyroid follicular epithelial cells.\(^11\)

These SCNs are multipotent cells which have been linked to several thyroid cancers, including papillary and MEC. Alternatively, it is thought that papillary carcinoma (the most common malignancy found in thyroglossal duct remnants) may undergo squamous and mucinous metaplasia resulting in MEC.\(^12\)\(^,\)\(^13\) However, it is still unclear that MEC in a thyroglossal duct remnant has arisen through the same means as a thyroid MEC or through another as yet unspecified mechanism.

Even though the treatment of thyroglossal duct carcinoma is still evolving, consensus have been attained in most areas. Conservative management with Sistrunk procedure alone is recommended for papillary thyroid cancer in low-risk patients who are female and younger than 40 years old, with no invasion of the capsule and a low-grade tumor of less than 1 cm in size.

In the case of large tumors >1 cm, invasion through the duct cyst wall, or suspect foci in the thyroid gland, a total thyroidectomy followed by I\(^131\) ablation and thyroid-stimulating hormone suppression is the most frequently proposed treatment.\(^14\) In pure squamous cell carcinomas of TG cyst, the Sistrunk procedure is the only warranted procedure.

Regarding MEC, the low-grade tumors can be treated by Cistrenk’s operation but aggressive treatment is warranted in high-grade tumors and the available literature supports the use of adjuvant radiotherapy in cases of high-grade tumors, which generally carry a poor prognosis.\(^15\) The use of radioiodine ablation due to the possible origins of these tumors having dedifferentiated from other thyroid neoplasms has been advocated by some authors.\(^16\)

Regarding cervical lymph nodes, even if these are not palpable, clinically frozen sections are considered to be mandatory. Radical or modified radical neck dissection is indicated only in the presence of positive lymph nodes.\(^17\)

As the frequency of nodal metastasis is relatively small the survival is 95.5% at 10 years.

**CONCLUSION**

We are reporting an usual histological surprise in a thyroglossal cyst malignancy being the only second reported case of TG cyst mucoepidermoid. This case highlights the importance of removal of thyroglossal duct cysts at an early stage and aggressive surgical approach in high-grade tumors.

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Work attributed to:
Government Medical College, Thrissur, Kerala, India

Orcid ID:
Ravindran Chirukandath - https://orcid.org/0000-0002-0562-1952
Nimisha CR - https://orcid.org/0000-0002-8530-4099
Babu PJ - https://orcid.org/0000-0003-1650-3609
Arya Puthukatt Muraleedharan - https://orcid.org/0000-0002-3431-0889
Lekshmi RS - https://orcid.org/0000-0001-7430-6903
Santhosh TV- https://orcid.org/0000-0001-7301-8269

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