An Unexpected Midline Neck Swelling in a Young Female

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An apparently healthy 26-year-old lady presented with a two-year history of painless upper neck swelling. There were no obstructive or constitutional symptoms. She denied features of hypo or hyperthyroid states and had none suggestive of obstructive or constitutional symptoms. She had no otologic nor nasal features.

On examination, the patient was comfortable in room air. Blood pressure: 110/70 mmHg, pulse rate 68 bpm, respiratory rate 16, afebrile. A 4 cm x 3 cm oval or multinodular firm, mobile (in all planes), non-tender infrahyoid midline mass, with normal overlying skin, was noted. The mass was well encapsulated and not adherent to surrounding structures with no external skin changes. Baseline blood investigations and thyroid function tests were within normal range. A flexible nasopharyngolaryoscopy performed revealed mobile vocal cords with normal supraglottic structures.

Cervical ultrasonography revealed a fluid-filled mass with solid components. Additionally, bilateral thyroid glands were present. Computed tomography neck revealed a septated complex cystic mass arising from the floor of the mouth measuring 2.9 x 3.1 x 3.3cm [Figure 1 (axial view) and Figure 2 (sagittal view)]. There was a focal and nodular enhancement of the septa measuring about 0.6cm, and the mass appeared to be continuous with the geniohyoid muscles. No cervical lymphadenopathy was noted.

Figure 1: Axial view of CT showing a septated complex cystic mass arising from the floor of the mouth measuring 2.9 x 3.1 x 3.3cm.
Figure 2: Sagittal view of CT showing septated complex cystic mass arising from the floor of the mouth measuring 2.9 × 3.1 × 3.3cm.

Question

What is your diagnosis?

a. Thyroid cyst
b. Thyroglossal duct Cyst
c. Thyroglossal duct carcinoma
d. Extrapulmonary tuberculosis

Answer

c. Thyroglossal duct carcinoma

A preliminary diagnosis of thyroglossal duct cyst was made. The patient was counselled for Sistrunk procedure. The surgery was successful, with clear margins and no lymphovascular invasion. There were no complications post-operatively, and the patient was discharged home on oral analgesia. The histopathological examination revealed papillary thyroid carcinoma of the thyroglossal duct cyst.

Thyroglossal duct cysts are remnants of the thyroglossal cyst that failed to involute between the 7th and 10th weeks of gestation.¹ These cysts can be found anywhere along the route of the thyroglossal duct and, depending on the rotation of the hyoid bone, can cause segmentation of the duct, whether in the pre-hyoid, transhyoid or retro hyoid position.¹ The thyroglossal duct can obliterate to form a fibrous cord or persists as a patent duct. A thyroglossal cyst is usually unilocular and can be isolated or associated with a duct or fibrous cord.²

Malignant thyroglossal cysts are rare.² Most cases are diagnosed post-operatively, but a diagnosis of malignancy is possible with FNAC.³ We decided to proceed with the Sistrunk procedure as the mass appeared to be arising from the thyroglossal tract. It is reported that the majority of thyroglossal duct carcinoma are Papillary Thyroid carcinoma (PTC). There is a predilection for females, with a 3:1 female-to-male ratio,⁴ and a mean age, at presentation, of about 40 years.⁵ Our patient was a female in her 20s, with an age that is suggestive of thyroglossal duct cyst.
The patient is currently on a two-monthly follow-up with a six-monthly imaging follow-up. Options of total thyroidectomy and RAI are still being discussed.

Thyroglossal duct carcinoma typically presents as an asymptomatic midline neck swelling, as reported in our patient. Nonetheless, features such as rapid expansion, fixed cyst and pain are tell-tale signs of malignancy of thyroglossal duct cyst. Rayess et al., reported that 73.3% of thyroglossal duct carcinoma are diagnosed incidentally, post-operative.

The ideal investigation for thyroglossal duct carcinoma is still debatable, although most authors favour imaging, either CT scan or ultrasound. The role of fine needle aspiration (FNA) remains questionable due to its low sensitivity, which is attributed to the dilution of the aspirate with cystic fluid contents.

It is worth noting that the presence of the thyroid gland must be confirmed before surgery. PTC of the thyroglossal duct may also occur as a primary entity due to ectopic thyroid gland tissue.

The Sistrunk procedure has been advocated by several authors as the majority of the diagnosis are typically made post-operatively. In patients diagnosed post-operatively with thyroglossal duct carcinoma, whether a total thyroidectomy solely or followed by radioiodine ablation is required remains debatable. Some authors propose that patients younger than 45 years with no lymph node involvement, metastatic spread, radiation exposure, or suspicious imaging findings with negative margins do not require routine total thyroidectomy. All patients with thyroglossal duct carcinoma with no suspicious metastatic spread require yearly imaging and blood investigations.

References