



# Papillary Thyroid Carcinoma Arising in a Branchial Cyst

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## OBJECTIVES

1. Describe the diagnostic dilemma of finding a thyroid carcinoma in a lateral neck mass without a primary.
2. Understand the etiological theories of branchial cleft cysts.
3. Adequately manage a patient with thyroid carcinoma in a lateral neck mass without a primary site identified.

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## INTRODUCTION

Branchial cleft cysts are the most common lateral cystic neck masses. Ectopic thyroid tissue within a branchial cleft cyst is a rare phenomenon and papillary thyroid carcinoma (PTC) arising from this tissue is extremely rare.<sup>1,2</sup>

We report a case of PTC incidentally found in a branchial cleft cyst. A total thyroidectomy and selective neck dissection were performed and no evidence of occult primary disease was found after review of fine sections. A brief discussion of the etiologies of these lateral neck masses is reviewed including how ectopic thyroid tissue may present in a lateral neck mass. Management recommendations regarding lateral neck malignancy without a primary is also presented.

## CASE

A 20-year-old man presented to the Otolaryngology Clinic with a history of painless right-sided neck swelling. Clinical examination showed a 6cm mass in his right neck without a cutaneous fistula. Contrast-enhanced CT scan and ultrasound of the neck confirmed a cystic lesion in the lateral neck without lymphadenopathy and a 2x4mm left thyroid nodule (figures 1 and 2). Surgical excision was performed and no tract was discovered on physical exam of the specimen during removal. Pathological examination found a 1cm focus of PTC within the 6cm cystic neck mass (figure 3). A second review of the specimen showed lymphocytic proliferation with PTC - no normal thyroid tissue was present within the branchial cleft cyst.

Thyroid function tests revealed a normal thyroid. After discussion with the patient a total thyroidectomy and selective neck dissection (central and ipsilateral) was performed presuming metastatic spread from a thyroid primary. Serial thin sections of the complete thyroid gland were analyzed and no carcinoma was detected. The subcentimeter nodule in the left thyroid lobe was found to be nodular hyperplasia. Twenty-one lymph nodes were included in the neck dissection and all were free of disease. To date, the patient's thyroglobulin levels have remained clinically negative.



Figure 1. Coronal view of a CT Scan with contrast showing right cystic neck mass. Lateral calcification corresponds to the area of malignancy.

## DISCUSSION

To our knowledge, only four cases have reported PTC arising in a branchial cyst without a primary in the thyroid.<sup>1-4</sup> At least a dozen cases of PTC in lateral neck cysts exist that either found an occult primary in the thyroid<sup>5-9</sup> or did not pathologically analyze the thyroid.<sup>10,11</sup> The presence of thyroid carcinoma in a lateral neck cyst is thought to be the result of metastatic spread. This is the obvious conclusion when a thyroid primary is found. However, we are left with a diagnostic dilemma in the absence of a primary – is this a case of metastatic disease with a missed primary carcinoma or rather PTC arising in ectopic thyroid tissue? If it is ectopic tissue in a lateral cyst, how did it get there?

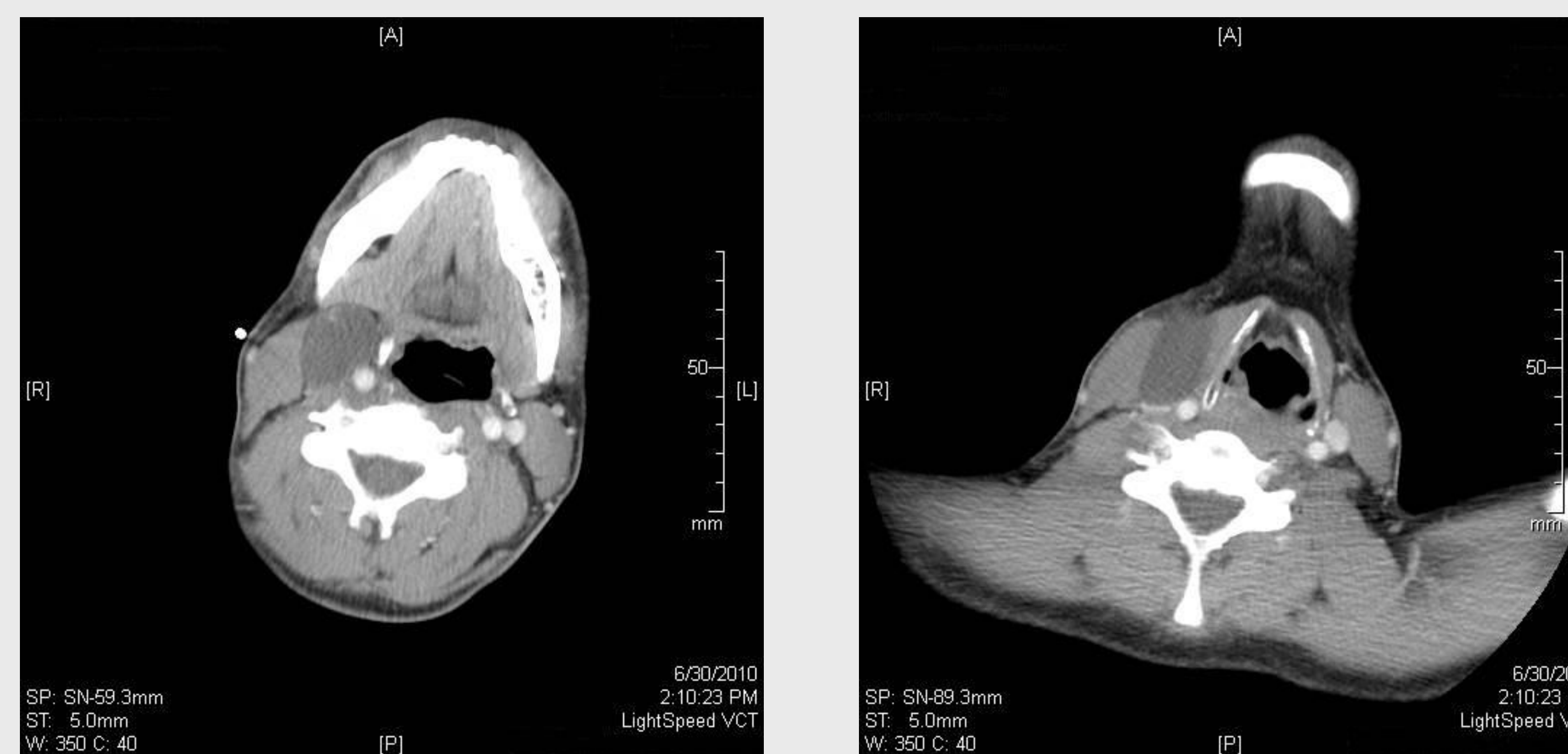


Figure 2. Axial CT Scans with contrast. Left – Cystic neck mass lateral to the great vessels. Right – compression of the jugular vein by the lateral neck mass is apparent.

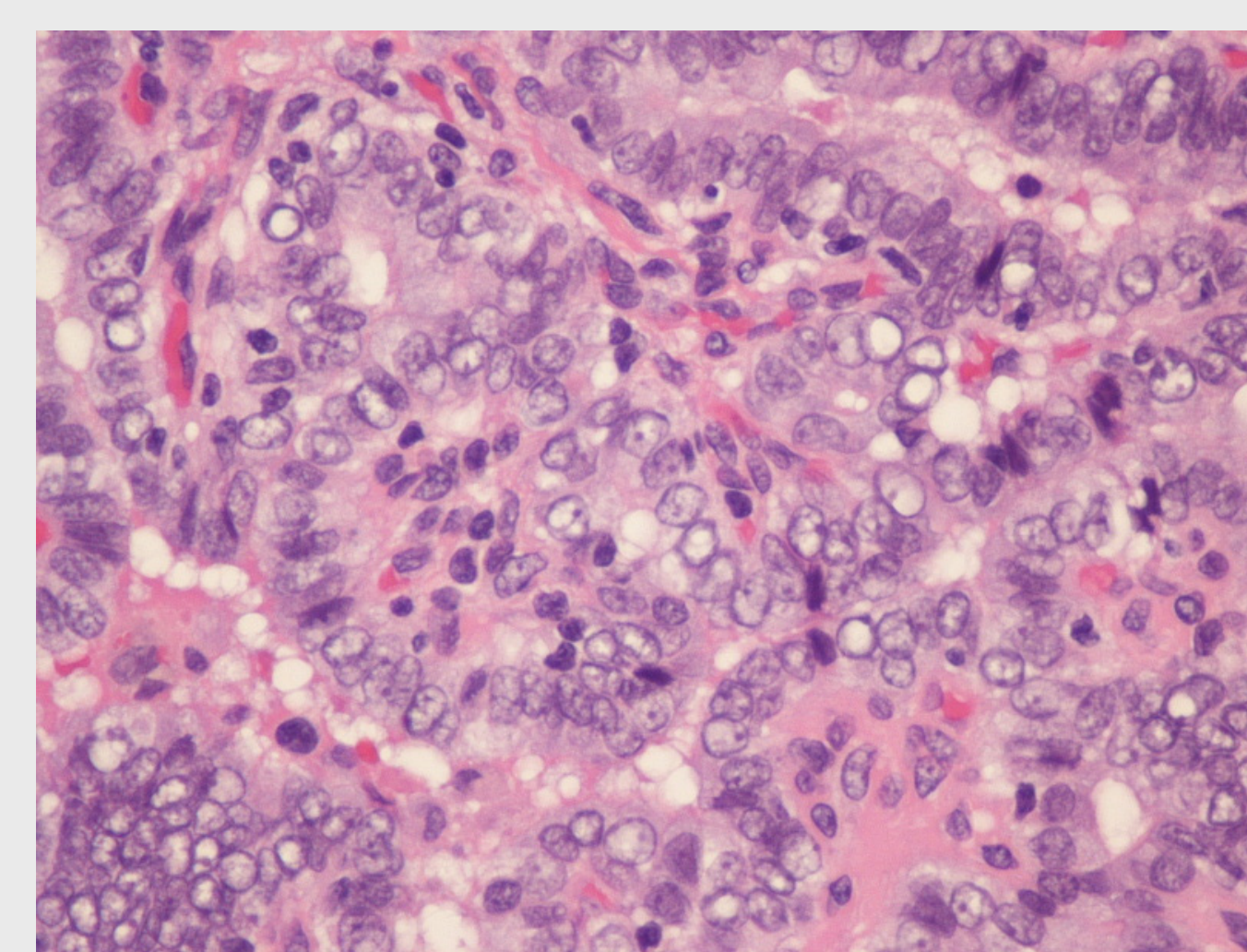


Figure 3. High power (40x) microscopy of lateral neck mass showing diagnostic clusters of papillary thyroid carcinoma.

Branchial cleft cysts are the most common lateral cystic neck lesion that typically present in the fourth decade of life<sup>6,12</sup>. However, despite considerable study of branchial cysts, its etiology remains to be fully elucidated. Older theories describe branchial cysts as congenital malformations resulting from failure of the branchial pouch apparatus. Recent theories, however suggest that some of these lateral neck masses are the result of cystic degeneration triggered by epithelial inclusions that migrate into lymph nodes.<sup>13-15</sup> Proponents of this acquired "inclusion theory" presence of lymph tissue<sup>17</sup> making the distinction between cystic lymph nodes and branchial cysts difficult. Some even propose using the term lateral or cervical lymphoepithelial cysts instead<sup>15,17</sup> since they appear to represent a separate etiology from true branchial cleft cysts that often have cutaneous or pharyngeal fistulas. This cystic change and de novo carcinoma could explain the pathophysiology in our case and has been proposed in similar reports<sup>1-4</sup>.

Another acquired theory proposes that the lateral anlage of the thyroid develops from the fourth-fifth branchial pouch and can aberrantly entrap normal thyroid tissue.<sup>16,18</sup> Thyroid development from a median anlage via the thyroglossal duct is undisputed. However, lateral contribution via the ultimobranchial body has been shown to contribute parafollicular C cells as well as thyroid glandular tissue.<sup>19-22</sup> Lateral branchial arch derivatives are implicated in contributing 1-30% of thyroid development.<sup>19,23</sup> These remnants may be termed thyrocarotid ducts<sup>23</sup> and may manifest as an arrest of lateral migration<sup>24</sup> or as the tubercle of Zuckerkandl. The proximity of branchial arches during development may also explain ectopic tissue (figure 4).

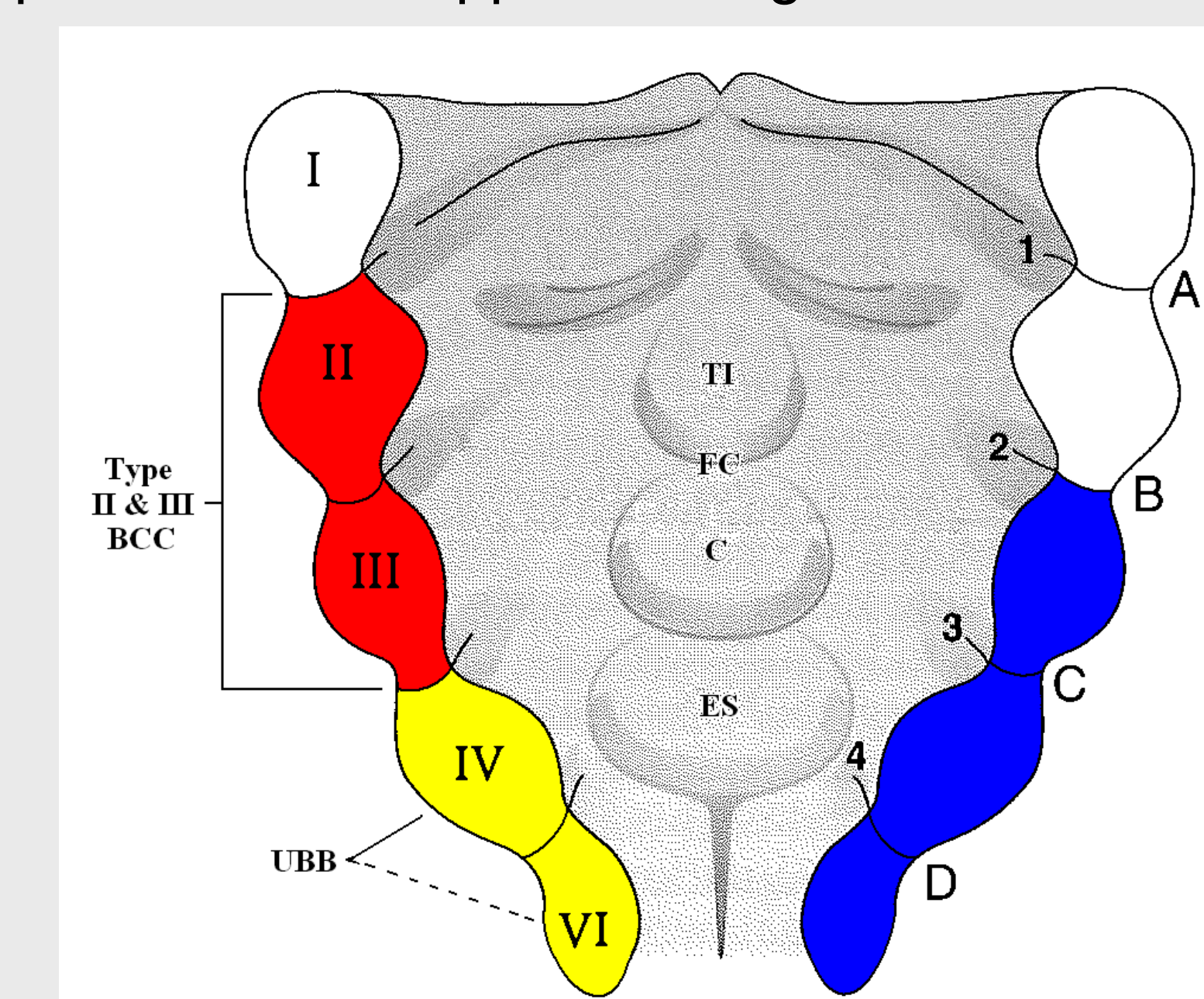


Figure 4. Coronal diagram of a 4-week fetus. I-IV branchial arches. 1-4 branchial pouches. A-D branchial grooves. Red section corresponds to areas of development associated with type II and III branchial cleft cysts. Yellow section corresponds to origin of suggested lateral thyroid anlage. Blue area corresponds to origin of superior and inferior parathyroid glands (4<sup>th</sup> and 3<sup>rd</sup> pouches respectively). TI = tuberculum impar; FC = foramen cecum; C = copula; ES = epiglottic swelling; UBB = ultimobranchial body.

## CONCLUSION

Regardless of etiology, the prognosis of patients with carcinoma in lateral neck cysts without a primary site identified after total thyroidectomy appears to be good.<sup>1-4</sup> This might suggest that removing the cyst is therapeutic if it represents de novo carcinoma in ectopic thyroid tissue. It could also represent a missed primary in the thyroid either secondary to a missed or "burnt out" microcarcinoma (occult carcinoma less than 1cm in diameter<sup>25</sup>) where a thyroidectomy may be considered appropriate treatment.

Although few examples exist of thyroid carcinomas presenting in a lateral neck cyst, management recommendations are similar in the literature. Imaging and fine needle aspiration cytology (FNAC) do not take the place of tissue diagnosis and excisional biopsy should be performed.<sup>6</sup> When the clinician is presented with a thyroid carcinoma in a lateral neck cyst a thorough examination of the neck is necessary. Imaging such as ultrasound and CT scan of the neck play a role to help distinguish the presence of metastatic nodes from branchial cysts as they can appear similar.<sup>5</sup> Total thyroidectomy is strongly recommended<sup>1-9,16,25</sup> and selective neck dissection should be considered.<sup>3,9,25</sup> Due to the possibility of papillary microcarcinoma, serial thin sections of all blocks of the totally embedded thyroid should be performed.<sup>3,7</sup> Adjuvant radioactive iodine might be considered if residual thyroid tissue or disease is suspected.<sup>9,25</sup>

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\*See handout for complete list of references.