



## GIANT PAPILLARY CARCINOMA THYROID- A RARE ENTITY

## ENT

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## ABSTRACT

**Background-** Papillary thyroid cancer (PTC) is the most common malignancy affecting the thyroid gland; many variants of PTC have been described. FVPTC is diagnosed on histopathology which shows a follicular architectural pattern with the typical nuclear features of the classical PTC (CPTC).

**Case presentation-** We report a case of 50 year old female patient case of giant follicular variant papillary thyroid carcinoma (FVPTC) with nodal & loco regional spread, who underwent total thyroidectomy with selective neck dissection followed by radioiodine therapy. Dimensions of the specimen were 17x 11x 8 cm and it weighed 1380 grams. Patient is on regular follow-up with no signs of any recurrent disease.

**Conclusion-** FVPTC's clinical behavior represents an intermediate entity with clinical features that are between classical papillary thyroid carcinoma and follicular thyroid carcinoma. Rarely, in some neglected cases FVPTC can attain enormous size, like in our patient's case the tumor weighed 1380 gms. Although these tumors vary in clinical behavior, long term outcomes are excellent and similar to classical papillary thyroid carcinoma.

## KEYWORDS

case report, follicular variant papillary thyroid carcinoma, thyroid tumor, giant thyroid tumor, neck swelling

## BACKGROUND-

Papillary thyroid cancer (PTC) is the most common malignancy affecting the thyroid gland; accounting for approximately 80% of all thyroid cancers. PTC typically occurs in middle aged with a peak incidence in 3<sup>rd</sup> & 4<sup>th</sup> decade with a female preponderance. Many variants of PTC have been described. Follicular variant of PTC (FVPTC) first described by Crile & Hazard in 1953 is the second most common form constituting between 9-22% of all PTC cases (1). FVPTC is diagnosed on histopathology which shows a follicular architectural pattern with the typical nuclear features of the classical PTC (CPTC) which include enlarged, overlapping or oval nuclei, nuclear clearings described as Orphan Annie eyes, nuclear grooves and pink cytoplasmic invaginations. Its clinical behavior represents an intermediate entity with features that are between CPTC and follicular thyroid carcinoma (FTC). Although these tumors vary in clinical behavior, long term outcomes are excellent and similar to CPTC. PTC is usually a slow growing neoplasm with early nodal metastasis. Early diagnosis & treatment reduces chances of distant metastasis. Even though thyroid swellings get noticed easily & usually present in initial stages, at times, neglected cases can assume enormous dimensions. We present a case report of giant FVPTC with nodal & loco regional spread, who underwent total thyroidectomy with selective neck dissection followed by radioiodine therapy.

## CASE PRESENTATION-

A 50 year old female patient was referred to our tertiary care center with a huge anterior neck swelling of 4 year duration with recent onset of breathing difficulty. She did not have any complaints of change in voice, difficulty in swallowing or any symptom suggestive of hypo/hyperthyroidism or distant metastasis. There was no history of any significant radiation exposure or any contributory family history. General condition of the patient was stable and there were no eye signs present. Examination of neck revealed a large 17 x 14 cm swelling over anterior aspect of neck with superior extension up to the level of hyoid,

inferiorly 2 finger widths below suprasternal notch over anterior chest wall. Laterally it was extending upto the posterior border of sternocleidomastoid on left side and upto anterior border of trapezius on right side (fig 1). Tumour had bosselated surface with stretched skin along with dilated veins and variegated consistency on palpation. Pemberton's sign was positive. Left level III cervical lymph node was found to be enlarged (3x3 cm). Indirect laryngoscopy was normal. Thyroid function test was within normal limits. Ultrasonogram of thyroid showed enlargement of both lobes with hypoechoic solid mass with irregular shape and cluster distribution of dot like calcification – falling to TIRADS 5 category. Contrast enhanced computed tomography scan of neck & chest showed a large heterogenous enhancing lesion of size 15x12x6 cm involving both lobes of thyroid gland with areas of necrosis and coarse calcifications (fig 2).

There was minimal retrosternal extension and mild tracheal compression. Left level III lymph node was enlarged with areas of central necrosis. Fine needle aspiration cytology showed high cellularity with clusters of follicular cells in focally prominent follicular pattern, suggestive of FVPTC.

After obtaining informed consent and preanaesthetic clearance, patient was taken up for surgery under general anesthesia. Total thyroidectomy with neck dissection (left II, III, IV, V & VI) was done (fig 3).

On left side the tumor was found adhered to recurrent laryngeal nerve which was shaved off from the nerve sheath. Laryngotracheal framework, esophagus and great vessels were not involved. Dimensions of the specimen were 17x 11x 8 cm and it weighed 1380 grams (fig 4). During the immediate postoperative period patient had left vocal cord paresis which recovered on conservative management. On histopathology, it was reported as follicular variant of papillary thyroid carcinoma with lymphovascular and capsular invasion. Level

II & III lymph nodes showed tumour deposits. Case was reviewed in tumor board and patient was advised I<sup>131</sup> scan followed by RAI. Now the patient is on regular follow-up with no signs of any recurrent disease.

## IMAGES



Figure 1- clinical picture of thyroid swelling



Figure 2- Thyroid gland with areas of necrosis and coarse calcifications



Figure 3- intraoperative image



Figure 4- postoperative specimen as described

## DISCUSSION

Papillary carcinoma and follicular carcinoma are the differentiated cancers of thyroid gland. Several variants of PTC have been described based on characteristic histological features. Common variants include follicular, classical and tall cell variant. Oncocytic, columnar cell, diffuse sclerosing & solid forms are some of the described uncommon variants.

In FVPTC, cells show follicular architecture, but their nuclear features show some of the typical features of PTC including ground glass appearance, nuclear pseudoinclusions, microfilaments and grooves.

FVPTC represents an intermediate entity with clinical features that are between C-PTC and FTC with respect to tumour size, extrathyroidal spread, lymph node and distant metastasis. The mean tumor size of FVPTC was slightly larger than that of C-PTC, but smaller than FTC. Extension into the extrathyroidal tissues and the involvement of lymph-nodes are more frequently found in FV-PTC than in FTC. Both these features which are important prognostic indicators are less common in FVPTC compared to C-PTC. Distant metastasis was less

common in FVPTC than in FTC, but more than CPTC (2).

Rarely, in some neglected cases FVPTC can attain enormous size, like in our patient the tumor attained a size of 17x 14 cm over 4 years. To the best of our knowledge, this is the largest case of FVPTC ever reported in literature. In a large study conducted by Xiao-Min Yu et al 10740 cases of FVPTC were reviewed from Surveillance, Epidemiology & End Result (SEER) Cancer Data base. Mean size of the tumour was 3.7 cm and the largest size was 5.9 cm (2).

Liu *et al.* classified FVPTC into two major classes: infiltrative (invasive) and encapsulated, which has a significant bearing on the outcome of disease profile (3). The infiltrative variant of FVPTC are more likely to have extrathyroidal extension and lymph node metastases. In our case also patient had these features of infiltrative variant. The encapsulated FVPTC clinically behaved more like follicular tumors.

There is lack of consensus on surgical management of FVPTC. Various studies recommend that for the encapsulated, noninvasive FVPTC, a lobectomy is enough and could decrease the chance of recurrence (4, 5, 6). In contrary, some authors advocates that the patients with FVPTC and PTC should receive the same treatment strategy (7,8). The American Thyroid Association (ATA) Management Guidelines recommends a total thyroidectomy if tumor measures > 4 cm, for low-risk carcinomas > 1 cm and < 4 cm, the treatment preferred is thyroid lobectomy, and tumors < 1 cm, lobectomy alone is preferred (9). In our case, owing to the large size of the tumour, we proceeded with total thyroidectomy with neck dissection.

According to ATA guidelines, definite candidates for radioiodine therapy includes tumors with gross extrathyroidal invasion (T4), pN1 with extranodal extension and ≥ 3 Lymph Node involved or any Lymph Node > 3 cm or patients with distant metastases. Our patient also received radioiodine therapy as per standard protocol and is on regular follow-up with no evidence of any recurrence.

Large thyroid swellings especially when malignant, it distorts the normal anatomy with displacement of structures making identification of recurrent laryngeal nerve difficult. When operating on these large thyroid tumors, there is increased risk of intraoperative bleeding leading to need for blood transfusion, increased risk of recurrent laryngeal palsy and overall morbidity.

## CONCLUSION

FVPTC's clinical behavior represents an intermediate entity with clinical features that are between classical papillary thyroid carcinoma and follicular thyroid carcinoma. Rarely, in some neglected cases FVPTC can attain enormous size, like in our patient's case the tumor weighed 1380 gms. In these large tumors, at-most care should be taken as there is increased chances of complication and overall increased morbidity. Although these tumors vary in clinical behavior, long term outcomes are excellent and similar to classical papillary thyroid carcinoma.

## Abbreviations-

PTC-	papillary thyroid carcinoma
CPTC-	classic papillary thyroid carcinoma
FVPTC-	follicular variant papillary thyroid carcinoma
RAI-	Radio Active Iodine
TIRADS-	thyroid imaging reporting and data system
ATA-	American thyroid association

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