Papillary Thyroid Carcinoma Presenting with a Right Preauricular and Intracranial Mass

ABSTRACT

Objective: To describe a case of a papillary thyroid carcinoma presenting with a preauricular and an intracranial mass and review the literature on the metastatic nature and invasiveness of papillary thyroid carcinoma.

Methods:
- **Design:** Case Report
- **Setting:** Tertiary Private Hospital
- **Patient:** One

Results: A 46-year-old female with a 12-year anterior neck mass and a two-year right pre-auricular pleomorphic adenoma on fine needle aspiration biopsy was found to have an intracranial mass on CT-scan. Total thyroidectomy and section biopsy of the preauricular mass yielded a final histopathologic report of follicular variant of papillary carcinoma, thyroid gland; and metastatic papillary thyroid carcinoma, follicular type, pre-auricular mass. The condition of the patient precluded neurosurgical intervention and RAI therapy and she underwent 23 sessions of external radiotherapy using 46Gy with significant diminution in size of the intracranial metastasis.

Conclusion: Papillary thyroid malignancy may be an indolent tumor but it is capable of distant metastasis. We should be alerted by host and tumor factors which can be predictors of a more radical papillary malignant disease whose management entails proper staging evaluation and good communication of prognostic data and available, realistic therapeutic options to patients using a multidisciplinary approach.

Keywords: papillary thyroid carcinoma; papillary thyroid carcinoma metastasis; infratemporal metastasis; brain metastasis of papillary carcinoma.

Thyroid malignancy is seventh among malignant lesions based on 1998 Philippine Department of Health statistics, of which Papillary thyroid carcinoma (PTC) is the most common, representing 82% of all thyroid cancers. PTC is believed to carry a good prognosis: overall mortality is 5% or less with long term follow up, with a survival rate for stage I and II disease (American Joint Committee on Cancer Staging) reaching up to 95% to 100%. PTC also has the lowest distant metastasis rates, ranging from 3% to 10%, with the majority of lesions affecting lungs and bone. While PTC is generally considered a malignant disease with a rather “benign” clinical character, surgeons should be aware of its dichotomous behavior. While some patients have excellent outcomes, others may surprisingly have a more aggressive disease.

This case reminds us of the clinically important dimorphism of PTC. The atypical dissemination to an unlikely anatomical region made this case interesting. Moreover, the remarkably silent yet amazingly extensive distant disease and manner of spread put to question the validity of common beliefs among regarding the clinical characteristics and behaviour of PTC. This paper will review available literature associated with remote spread of disease even when clinical warning signs are
lacking, and explore plausible mechanisms for the pathogenesis of the extraordinary dissemination observed in this case.

**CASE REPORT**

A 46-year-old female consulted for a gradually-enlarging anterior neck mass of 12 years duration and a 2-year right pre-auricular mass. Examination revealed a 7.0 cm x 5.0 cm anterior neck mass that was firm, non-tender and moved with deglutition. There were no palpable cervical lymph nodes. The firm, non-movable and non-tender pre-auricular mass measured 5.0 cm x 5.0 cm (Figure 1). Thyroid function tests were normal. Fine needle aspiration biopsy (FNAB) of the anterior neck mass was read as follicular neoplasm, while that of the pre-auricular mass was signed out as consistent with pleomorphic adenoma.

A computed tomography (CT) scan revealed a large enhancing lobulated soft tissue mass in the right pre-auricular area measuring 5.9 cm. x 6.33 cm. x 6.79 cm. (Figures 2 a, b). Superiorly, the mass extended into the right temporal lobe associated with lytic destruction of the squamous and mastoid portions of the temporal bone. Inferior extension reached the level of the mandibular ramus. The masseter appeared infiltrated. There was erosive destruction of the zygomatic arch, greater sphenoid wing, right anterior skull base and anterior wall of the glenoid fossa of the temporomandibular joint. Posteriorly, there were erosive changes along the anterior wall of the mastoid bone. The lesion abutted the anterior surface of the parotid gland without infiltration. Another similarly enhancing lesion was noted on the midoccipital region, measuring 2.96 cm. x 3.66 cm with extension into the posterior fossa, adjacent to the confluence of sinuses, associated with right anterolateral displacement of the torcula of Herophilii and lytic occipital bone destruction. The right lobe of the thyroid gland was enlarged measuring approximately 6.0 cm. x 3.6 cm. x 2.7 cm. There were hypodense lesions seen in the right lobe, some with dense peripheral calcifications; the largest measuring 3.4 cm. x 3.1 cm. x 2.2 cm. The left lobe was normal in size with inhomogenous density and a focus of calcification.

A total thyroidectomy and section biopsy of the preauricular mass were performed. Final histopathologic report for the thyroid gland lesion was follicular variant of papillary carcinoma with note of lymphatic and vascular invasion and thyroid capsular spread. The pre-auricular mass was signed out as consistent with metastatic carcinoma, follicular pattern, thyroid in origin. The final diagnosis was papillary thyroid carcinoma, right and left thyroid gland with brain (temporal and occipital lobes) metastases; T4aN0M1; Stage IVc (AJCC Classification, 2002).

Total body scans revealed functioning thyroid tissue remnants in the anterior neck, with no evidence of distant thyroid cancer metastasis (Figure 3). The patient was subjected to 23 sessions of external beam radiotherapy (EBRT) at 46Gy per session with focus on the right pre auricular region and mid occipital area. After one month of EBRT, the pre-auricular and occipital mass had significantly decreased in size (Figures 4 a, b).

**DISCUSSION**

The presence of distant metastasis from PTC is unusual. Dinneen et al. described the distribution of distant metastasis from PTC among a hundred cases during a five decade period. The most commonly involved organs were the lungs (77%) and bone (20%). Other sites include the mediastinum (10%), adrenals (1%), skin (1%) and liver (1%). The brain is only involved in 1% of cases. Despite multiple large intracranial masses, this patient did not have any sign or symptom of increased intracranial pressure nor focal neurologic sign on examination.

In a study by Chiu et al. among PTC patients with brain metastasis, 23% of the study population (11 out of 47 patients) also had no clinical evidence of distant brain metastasis, with most only diagnosed post mortem. This was probably because the mass usually lysed cranial bones, the “auto-cranietomy” dissipating intracranial pressure into the more distensible scalp tissue. Most patients with clinically evident brain metastasis were those who developed deeply-seated brain lesions with practically no calvarial bone involvement, thereby increasing intracranial pressure.

The mechanism of distant spread of papillary thyroid carcinoma has been postulated to be due mainly to lymphatic and vascular invasion by the tumor, following pre-formed neural and lymphovascular anatomical pathways through natural foramina, fissures and hollow fascial spaces.

The mean time for development of distant metastasis was noted to be around seven to eight years after diagnosis of early stage disease. In our case, it took 10 years for distant metastasis to be evident. Dineen showed that age at the time of distant metastasis diagnosis was a strong predictor of PTC survival after distant metastasis with a 10-year survival rate of 82% for those less than 40 years of age compared with only 18% for the group aged 65 years and older at the time of distant metastasis. Gender did not significantly influence PTC survival after distant metastasis. Distant metastasis in other organs in the body may be related to brain metastasis; Dineen’s study showed that in patients who were positive for distant organ involvement, the brain had the highest risk for development for a second organ or succeeding metastases. Our patient only had brain metastasis without the more common involvement of lung and bone.

The predilection of PTC to spread through the lymphatic more than the hematogenous route has been well established. Hall showed that intratumoral lymphatic proliferation and invasion of new lymphatics in and around the enlarging malignant tumor may facilitate distant spread. Moreover, enormously large loads of malignant cells in the lymphatics may cause indirect vascular invasion as lymphatic channels ultimately drain into vascular channels. The intratumoral lymphatic invasion seen in our histologic specimen may explain its aggressiveness.
CASE REPORTS

Figure 1. Preauricular Mass. Patient had a right pre-auricular mass measuring approximately 5x5cm.

Figure 2a. Contrast CT scan of Head, Axial Section showed an enhancing lobulated soft tissue mass, involving right masseter and temporalis muscles, measuring 5.99 x 6.37 x 6.79cm.

Figure 2b. Contrast CT scan of Head, Coronal Section showed an enhancing soft tissue mass measuring 5.99 x 6.37 x 6.79 cm, extending superiorly to the right temporal lobe and inferiorly to the level of the mandibular ramus.

Figure 3. Total Body Scan (I-131). This showed intense tracer uptake in the anterior neck. Pinhole imaging in this area shows 3 foci of increased tracer deposition in midline measuring 1.6x1.1cm, 1.2x2.0cm, 1.1x1.2cm as well as a faint focus in the superior aspect. There is also a focus of tracer localization in the right thyroidal bed measuring 1.0x1.0cm. Functioning thyroid tissue remnants in the anterior neck. No evidence of distant thyroid cancer metastasis.
intrathyroidal evidence of vascular invasion as predictive of disease recurrence among their patient population. This mode of malignant dissemination may better explain the intracranial metastasis in our case in the absence of clinical cervical node involvement.

Another factor which may explain the aggressiveness of the papillary thyroid carcinoma in this case was the extracapsular invasion. According to Machens et al, extrathyroidal growth consistently emerged as the most significant factor for increased likelihood of distant metastasis in PTC. Extracapsular and extrathyroidal spread give the malignant cells more avenues for lymphatic and vascular dissemination through adjacent tracheal, strap muscles, nerve tissue and other contiguous structures.

The notion of follicular variant of papillary thyroid carcinoma (FVPTC) having a more aggressive clinical nature than the pure type of PTC has been disproved. Even with a predominant follicular component, lesions with papillary features behave clinically as true papillary carcinoma. A clinico-pathologic analysis of 243 patients with pure versus follicular variant papillary thyroid carcinoma by Jamal et al showed that pathologic and clinical behaviours of PTC and FVPTC were comparable. Prognostic factors, treatment and survival also were similar between the two forms. The follicular variant seen in our patient may therefore not relate as much to tumor aggressiveness.

While the usual therapeutic modality for high-risk papillary thyroid carcinoma is total thyroidectomy with radioactive iodine (RAI) ablation, the management of brain metastasis may relate more to palliation than cure. Although surgical resection has been shown to prolong survival by nearly three-fold in 47 cases of thyroid malignancy with brain metastasis, our neurosurgery service advised against surgery due to multifocal brain involvement, uncertainty of complete resection and absence of neurological deficit.

Initial radiation therapy prior to RAI ablation was advised because the intracranial lesion was in very close proximity to the venous confluence of sinuses which could swell with RAI and result in massive cerebral edema. Radiotherapy was probably a reasonable option as previously recommended for patients unable to undergo surgical therapy and who had disseminated or inaccessible intracranial lesions that precluded metastasectomy. Our patient underwent 23 sessions of EBRT using 46Gy with subsequent remarkable decrease in the size of the brain metastatic lesions. As of this writing, she was asymptomatic and awaiting possible RAI therapy to further decrease the size of the intracranial lesions.

In summary, while brain metastasis from a PTC is unusual, several features of the host (older age, long duration of disease and presence of distant metastasis) and the tumor pathology (presence of intratumoral lymphatics, vascular invasion and extrathyroidal invasion) may alert the clinician to the risk for intracranial spread and possible need for a cranial CT or MRI. While the prognosis may be dismal, surgical resection offered a significant longer survival for most patients in the literature.

The aggressiveness of PTC in this case may be further related to actual vascular invasion demonstrated in our thyroid specimen. Vascular invasion in PTC occurs in only 2 to 14% of cases. Gardner et al. highly correlated vascular invasion, whether intrathyroidal or extrathyroidal, with more aggressive local disease and a greater predisposition for distant metastasis at diagnosis. Their study also identified intrathyroidal evidence of vascular invasion as predictive of disease recurrence among their patient population. This mode of malignant dissemination may better explain the intracranial metastasis in our case in the absence of clinical cervical node involvement.

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Other palliative management such as RAI ablation and radiotherapy remain as possible options should the situation preclude surgical intervention.

This case study serves to remind otolaryngologists to be more vigilant in the diagnosis and management of patients with PTC. Increasing numbers of PTC cases in our institution are displaying more aggressive behaviour. Their management entails proper staging evaluation and good communication of prognostic data and available, realistic therapeutic options to patients using a multidisciplinary approach.

REFERENCES

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