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Case Report

Diffuse Sclerosing Variant of Papillary Thyroid Carcinoma Presenting with Brain Metastases

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Submitted: 21 July 2016

Accepted: 08 September 2016

Published: 09 September 2016

Keywords

- Papillary thyroid carcinoma
- Metastases
- Aphasia

Abstract

Diffuse Sclerosing Variant of Papillary Carcinoma of the thyroid (DSVPC) is a rare variant of Papillary Thyroid Cancer (PTC). It is characterized by diffuse involvement of one or both thyroid lobes, with dense fibrosis, squamous metaplasia, and lymphocytic infiltration. It has more aggressive course, and higher rate of metastasis compared to the conventional PTC. We describe a female with DSVPC who presented with hemorrhagic brain metastasis, and pulmonary metastasis.

CLINICAL HISTORY

65-year-old female presented with acute onset expressive aphasia. Past medical history was significant for goiter. On review of systems, she reported 40 pounds weight loss over the last year, and cough for the last 2-3 weeks. Except for expressive aphasia and mildly dysarthric speech, her physical exam was unremarkable.

WORKUP

MRI of the brain was done. This showed 4 small enhancing lesions in the left cerebral hemisphere. One of the lesions was hemorrhagic (Figure 1). CT chest abdomen pelvis was obtained to evaluate for a primary source of malignancy. This revealed a diffusely enlarged and heterogeneous thyroid gland, with mass effect and tracheal deviation (Figure 2a). In addition, it showed supra clavicular and mediastinal lymphadenopathy, and multiple metastatic lung masses (Figure 2b). Next, thyroid US was performed. This showed diffusely enlarged thyroid gland with micro calcifications also showed diffuse cervical lymphadenopathy with micro calcifications (Figure 3a,3b, 3c,3d). Fine needle aspiration cytology (FNAC) of the cervical lymphadenopathy was positive for papillary thyroid cancer. The brain and lung metastases were presumed to be most likely due to metastatic papillary thyroid cancer.

DISCUSSION

Diffuse sclerosing variant of papillary carcinoma of the thyroid (DSVPC) is rare variant of papillary thyroid cancer (PTC). On sonography, it is characterized by diffusely enlarged thyroid gland with heterogeneous hypoechogenicity and diffuse scattered micro calcifications, resulting in "snowstorm

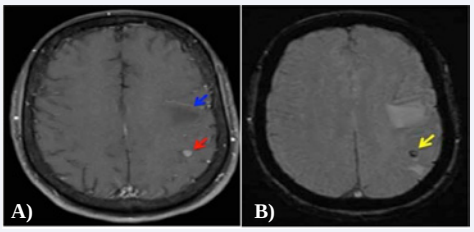

Figure 1 (a) Axial postcontrast T1FS of the brain shows a small enhancing focus in the left parietal lobe (red arrow). Anterior to the lesion is an area of T1 prolongation (blue arrow), consistent with vasogenic edema associated with a different metastatic lesion (not shown on this slice). (b) Susceptibility Weighted Imaging (SWI) demonstrates blooming artifact at the enhancing lesion (yellow arrow), consistent with a hemorrhagic metastasis.

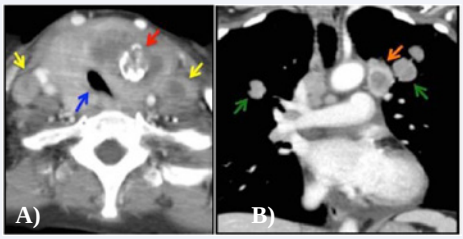

Figure 2 Contrast enhanced CT. (a) Axial image through the lower neck demonstrates a diffusely enlarged and heterogeneous thyroid gland, left lobe more than the right. There is a heavily calcified left thyroid nodule surrounded by an area of hypo attenuation, likely necrosis (red arrow). There is mass effect on the trachea, which is deviated to the right (blue arrow). Also there is bilateral supraclavicular lymphadenopathy, necrotic on the left (yellow arrow). (b) Coronal image of the chest shows bilateral lung masses (green arrow) and mediastinal lymphadenopathy (orange arrow).

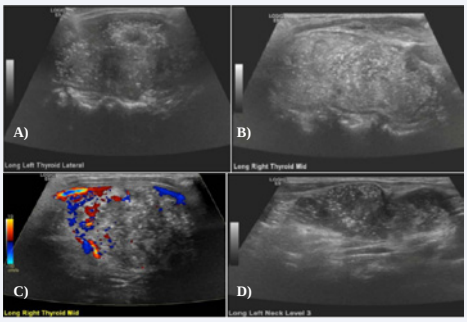

Figure 3 Thyroid and neck US (a) (b) Right and left thyroid lobes are diffusely enlarged and heterogeneous with diffuse micro calcifications. (c) Color Doppler US of the right thyroid lobe shows increased intrinsic vascularity.

(d) Left level III cervical lymph node is enlarged with lobulated contour, diffuse micro calcifications, predominantly hypoechoic background, and absent fatty hilum.

appearance" [1,2]. Histopathologically, DSVPC is characterized by diffuse involvement of one or both thyroid lobes, with dense fibrosis, squamous metaplasia, lymphocytic infiltration, psammoma bodies, and areas of conventional papillary cancer [3]. The incidence of DSVPC ranges from 0.75 to 5.3% [4].It has higher prevalence in younger women, larger tumor size, and higher incidence of cervical lymph node metastases, and pulmonary metastases [3,5].DSVPC is frequently associated with Thyroglobulin antibodies (75%) [4]. The combination of this finding and the diffuse involvement of the thyroid gland can be confused with Hashimoto's thyroiditis, and can possibly result in delayed diagnosis. The combination of diffusely enlarged and heterogeneous thyroid gland with diffuse scattered micro calcifications and cytopathology result of papillary thyroid cancer should raise the suspicion for DSVPC.CT or MRI can be used to evaluate the extra thyroidal extent of disease. Iodine 131 whole body scan is used to evaluate for metastases or recurrent disease. Fluorodeoxy glucose positron emission tomography (FDG PET) can also be used for staging, particularly in patients with non-iodine-avid disease [6]. The distant metastasis rate in DSVPC is up 28%, compared to 10-12% in classical PTC [4,6,7]. Pulmonary metastases are the most common distant metastases with PTC (80%) [7]. Metastases to the brain are extremely rare (1.2%), and tend to be hemorrhagic and have very poor prognosis [7]. Overall, PTC has good prognosis with 5-year survival up to 98% [6]. However, the prognosis of DSVPC is controversial. Some authors report less favorable prognosis due to the extensive nature, and higher incidence of cervical lymph node metastases, with or without distant metastases. However, more recent studies reported similar prognosis to the classical papillary thyroid cancer, due to young age, aggressive treatment, and close monitoring [2,4]. The definitive treatment of papillary thyroid cancer, including DSVPC is surgery. This is usually followed by radioiodine scintigraphy to detect residual or metastatic disease, which is treated by therapeutic Iodine 131. Adjuvant radiotherapy is sometimes used, especially in patients older than 45 years with local invasion. Patients will require lifelong treatment with thyroid hormone replacement after total thyroidectomy.

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Cite this article

Amarneh M, Abu-Yousef M (2016) Diffuse Sclerosing Variant of Papillary Thyroid Carcinoma Presenting with Brain Metastases. JSM Clin Med Imaging Cases Rev 1(1): 1005.