Long-Term Follow-Up of a Primary Teratoma With Somatic-Type Malignancy Within the Thyroid Gland Mimicking Thyroid Carcinoma

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Clinical Practice Points

- Primary teratoma of the thyroid as an extragonadal germ cell tumor (EGCT) in men is a very rare disease with highly malignant potential and poor prognosis. As a secondary tumor, EGCT after burned out gonadal tumor leaving a fibrous scar can mimic a primary EGCT; accurate staging procedures are always mandatory.
- This report presents the first case of EGCT in an adult male thyroid gland with long-term disease-free survival (12 years since salvage chemotherapy).

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Introduction

Primary extragonadal localization of germ cell tumor (GCT) is a very rare disease found in the midline of the body, predominantly in anterior mediastinal (50%-70%) or retroperitoneal (30%-40%) areas.[1] Whereas most teratomas involving the thyroid are benign and occur in children, the adult cases are rare and mostly malignant,[2] with one exception of recurrent benign teratoma reported by Keynes in 1959.[3] To date, 2 fatal cases of primary teratoma within the thyroid in men have been described in the literature[4,5]; the present report is the first describing a long-term disease-free survival of 12 years after salvage chemotherapy.

Case Report

A previously healthy 43-year-old man presented with a 2-month history of enlarged left portion of the thyroid gland without narrowing of the trachea on radiologic evaluation. On palpation, the mass was solid, and the results of preliminary laboratory tests, including thyroid function (free triiodothyronine, free thyroxin, thyroglobulin, and thyrotropin), carcinoembryonic antigen, and prostate-specific antigen tests, were within the reference ranges. Ultrasonography found a mass of 5 × 3.5 cm within the left lobe of the thyroid gland, and there was no accumulation of technetium-99m pertechnetate on thyroid scintigraphy (cold thyroid nodule). Fine-needle aspiration of the mass was performed, and cytology found atypical cells probably belonging to a malignant tumor. Hence, frozen section–guided surgery was scheduled. Preliminary histology found a teratoma with somatic-type malignancy, consisting of a primitive neuroectodermal tumor (PNET) (Figure 1). Consecutively, a left unilateral lobectomy and a subtotal resection of the right thyroid lobe were performed. On the left side, small parts of the pseudocapsule surrounding the tumor could not be completely removed from the carotid artery. Postoperative levels of α-fetoprotein and β-human chorionic gonadotropin were within reference ranges. Lactate dehydrogenase was within reference ranges when first analyzed on postoperative day 20, after the patient had been referred to the authors' department. Final histology confirmed the diagnosis; histologically, the tumor margins were negative. Staging computed tomography (CT) scans of the head, neck, thorax, abdomen, and pelvis were negative for disease. Despite normal findings from physical examination of the scrotum and scrotal sonography with 15 Hz, testicular biopsies were performed for assessment of testicular intraepithelial neoplasia (TIN); immunohistochemistry did not provide any evidence of malignant or premalignant cells. Tumor marker results remained negative during the restaging period. Finally, 1 month after primary surgery, 3 adjuvant cycles of PEB (consisting of cisplatin, 20 mg/m^2 on days 1 to 5; etoposide, 100 mg/m^2 on days 1 to 5; and bleomycin, 30 mg on days 1, 8, and 15) were administered to the patient because of

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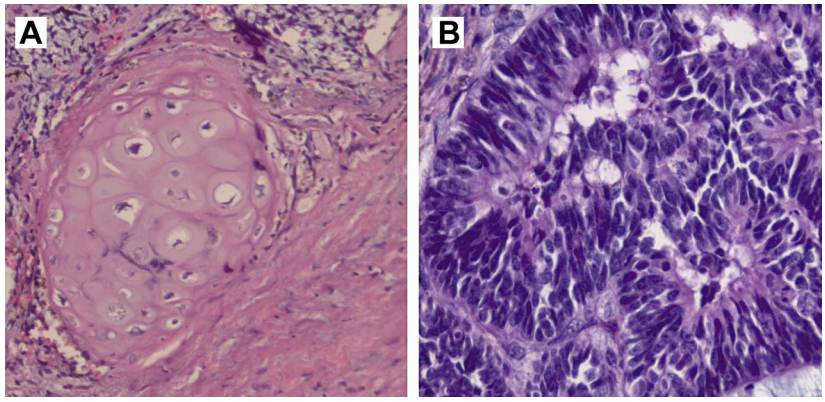
Figure 1 Teratoma With Cartilage (A) and Somatic-Type Malignancy Consisting of Primitive Neuroectodermal Tumor Forming Rosettes and Ribbons (B)



macroscopic possible positive margins and the reduced long-term outcomes of patients with extragonadal germ cell tumor (EGCT) compared with those with tumors of gonadal origins.[1] After 24 months of follow-up, enlarged lymphatic nodes were noticed at the left sternocleidomastoid muscle, and CT scan found enlarged lymph nodes and a tumor mass extending to the anterosuperior mediastinum (Figure 2). Recurrence was confirmed by biopsy, and 4 cycles of PEI (cisplatin/etoposide/ifosfamide) were subsequently administered. At 6 weeks after chemotherapy, complete response was observed on CT scan. Extended left-sided neck dissection and sternotomy were performed to clear all lymphatic tissue (so as to detect any active tumor and to remove teratoma) in the area of the tumor before chemotherapy. In meticulous histologic workup, extensive necrosis and regression with large numbers of macrophages and scar formation were present, as well as small parts of the teratoma with somatic-type malignancy (PNET). The patient was free of disease at the time of this report, 12 years after salvage chemotherapy.

Discussion

GCT is the most common neoplasm in young men, generally occurring in the third or fourth decade of life.[6] Generally, the origin is in the gonad; only 2% to 5% of all GCTs occur primarily in the mediastinum, retroperitoneum, thymus, sacrococcyx, prostate, stomach, pineal gland, and some other unusual sources.[1,7] Also, a secondary EGCT after burned out gonadal tumor leaving a fibrous scar may mimic a primary EGCT; therefore, accurate staging procedures are mandatory in terms of detecting a possible small testicular primary tumor or especially TIN not necessarily eradicated by chemotherapy.[6,8] Reduced response of TIN to chemotherapy

Figure 2 At 24 Months After 3 Adjuvant Cycles of Cisplatin/Etoposide/Bleomycin: Lymph Node Metastasis at the Left Sternocleidomastoid Muscle Extending to the Anterosuperior Mediastinum