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Case Report

Malignant epithelioid hemangioendothelioma of the thyroid: A case report

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Primary malignant epithelioid hemangioendothelioma (MEH) of the thyroid is a rare neoplasia with only a few cases reported in the literature. We report a 75 year old man, who presented with a substernal goiter and compressive symptoms. Ultrasonographic evaluation revealed a hypoechogenic nodule in the left lobe, measuring 4.1 cm in maximum diameter, and associated gross calcifications. Fine needle aspiration yielded hemorrhagic material. A left thyroid lobectomy and isthmusectomy was performed. The surgical specimen contained a malignant epithelioid hemangioendothelioma measuring 6 x 4 x 3 cm that had infiltrated about 50% of the thyroidal parenchyma, and surrounded a necrotic nodule. Immunohistochemistry results corroborated the histopathological findings; staining was positive for AE1/AE3, CD31, CD34, factor VIII-related antigen, and Ki-67 expression. Because of the patient's comorbidities, surgical complementation was not undertaken and he has been undergoing conservative treatment.

Key words: Thyroid nodules, hemangioendothelioma, goiter, rare neoplasia.

INTRODUCTION

Thyroid nodules are a very common finding in daily clinical practice, and are the principal manifestation of several thyroid diseases. The most common causes are colloid cysts and thyroiditis (80% of cases), followed by benign follicular neoplasias (10 to 15%) and carcinomas (5%) (Coltrera, 2008; Gharib and Papine, 2007; Hegedüs et al., 2003).

Vascular tumors of the thyroid include benign hemangiomas and, rarely, hemangiendotheliomas and angiosarcomas, which have been reported principally from the Alps region of central Europe, where iodine deficiency is endemic. Malignant epithelioid hemangioendothelioma (MEH) has histological features that are intermediate between those of benign hemangiomas and conventional angiosarcomas.

Histologically, MEHs are classified into the following 4 categories: epithelioid, spindle cell, kaposiform hemangioendothelioma, and malignant papillary endovascular endothelioma (Egloff, 1983).

MEH is a rare tumor that occurs in soft tissues, the lungs, liver, bones, and, even more rarely, in the thyroid and pleura (Crotty et al., 2000). In western Austria, the incidence is estimated to be 0.15 to 0.25 cases per 100,000 inhabitants per year (Rhomberg et al., 2004). The objective of this report is to present the case of a 75 year old man with substernal goiter, in whom MEH of the thyroid was diagnosed.

CASE REPORT

A 75 year old man with a 12 year history of coronary artery disease was referred to our service for investigation of a cervical nodule that had been present for 3 years, accompanied by compressive symptoms. The reported complaints included dysphagia for solid food and slight discomfort in the cervical region. The

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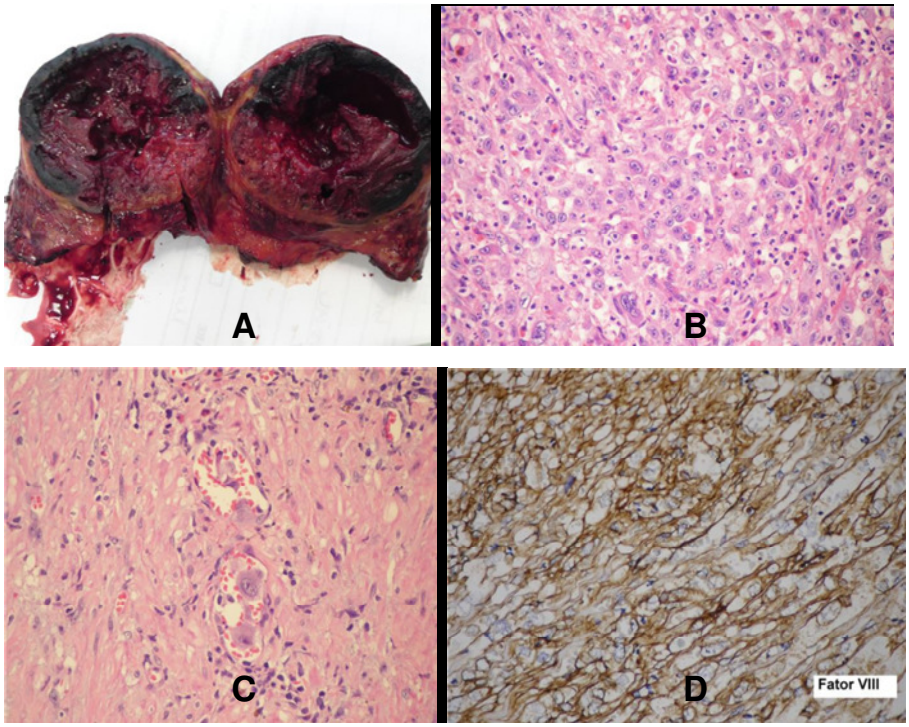

Figure 1. Malignant epithelioid hemangioendothelioma of the thyroid. A: macroscopic lesion (thyroid lobe surface appears wine-like and smooth, with bright aspects; weight 69 g; and measuring 7 × 4 × 3 cm); B and C: histopathology (infiltrative neoplasia characterized by vascular channels made up of polygonal pleomorphic cells with high nucleus-to-cytoplasm ratios and condensed chromatin, and an elevated mitotic index); D: immunohistochemical staining positive for factor VIII-related antigen.

patient had a history of tobacco smoking, myocardial revascularization surgery (in 1999), and 1 episode of spontaneous pneumothorax of unknown etiology. He was being monitored by a urologist for ureter stenosis, low-grade papillary urothelial carcinoma, and benign hyperplasia of the prostate. He had no history of prior radiation. Physical examination revealed a firm cervical nodule in the region of the thyroid, predominantly on the left side; it was mobile on deglutition and measured approximately 4 cm. There were no palpable lymph nodes, and the rest of the clinical examination was unremarkable.

Ultrasonography revealed a hypoechogenic nodule in the left lobe of the thyroid that was associated with gross calcifications and measured 4.1 cm in its greatest diameter. Fine-needle aspiration biopsy (FNAB) initially only yielded amorphous material, suggestive of colloid. A repeat biopsy yielded hemorrhagic material. Thyroid function was within normal limits. The patient was referred for surgical treatment because of worsening compressive symptoms. A left thyroid lobectomy and isthmusectomy were performed. Examination of the resected tissue revealed a malignant epithelioid hemangioendothelioma measuring 6 × 4 × 3 cm that had infiltrated about 50% of the thyroidal parenchyma, and surrounding a necrotic nodule. Approximately 2 mitoses/10 high-power fields and discrete intramural lymphomononuclear inflammatory infiltrates were noted. Immunohistochemistry results corroborated the histological findings; staining was positive for AE1/AE3, CD31, CD34, factor VIII-related antigen, and Ki-67 expression (Figure 1). Because of the patient's comorbidities, principally cardiac disease, surgical complementation was not undertaken. The patient was monitored by specialists in endocrinology, metabology, head-and-neck surgery, and oncology. He received cervical radiotherapy at a total dose of 4000 cGy. One year after his partial thyroidectomy, the patient presented consumptive syndrome and evidence of possible tumor recurrence in the lungs.

DISCUSSION

The first case of thyroid MEH was described in 1953 by Bacher in a report of a highly malignant thyroid tumor (Bacher, 1953). The term epithelioid hemangioendothelioma was first used in 1982 by Enzinger and Weiss (Enzinger, 1995) for a group of vascular tumors found in such tissues as skin, bone, lung, pleura, liver,

peritoneum, and mediastinum (Yousem and Hochholzer, 1987; Suster et al., 1994). At one time, differentiation between MEH and angiosarcoma was difficult, which might have contributed to conflicting reports (Van Beers et al., 1992). However, small nests and cords and fusiform cells with intracytoplasmic vacuolization are typical of MEHs and generally cannot be observed in angiosarcomas. Grossly, MEH usually occurs in the form of a solid tumor that may contain areas of hemorrhage and necrosis (Weiss et al., 1986; Mills et al., 1994).

In the MEH specimen from this case, the commonly expressed markers included AE1/AE3, CD31, CD34, factor VIII-related antigen, and Ki-67.Ki-67 indicates a high level of cell proliferation, while factor VIII-related antigen and CD31 are markers seen in vascular tissues. The diagnosis of thyroidal MEH is generally only made by surgical exploration of the neck, because results of laboratory tests and imaging exams are nonspecific. Occasionally, in experienced hands, FNAB can provide the diagnosis (Siddiqui et al., 1998; Luze et al., 1990), although the hemorrhagic characteristics of the tumor may be an impediment. The symptoms of the disease are often nonspecific and are very similar to those of anaplastic carcinoma, except that distant metastases in MEH are frequently hemorrhagic (Weiss et al., 1986; Mills et al., 1994). Patients usually only live a few months after the diagnosis, and profound anemia and striking cachexia are common. There has been little clinical evidence of positive response to surgical treatment, radiotherapy, or chemotherapy (Egloff, 1983; Thaler et al., 1986). Rhomberg et al. (2004) reported on 12 patients, of whom 5 had unexpectedly favorable outcomes.

Our patient with MEH of the thyroid presented with evidence of metastases to the lungs in chest tomography 1 year after partial thyroidectomy. During his initial treatment, he had undergone radiotherapy as adjuvant therapy after surgical excision, despite the paucity of evidence for positive outcomes using this approach. In summary, MEH of the thyroid is a rare tumor that can be extremely aggressive. It is found principally in the Alps of central Europe, where iodine deficiency is endemic. In experienced hands, a diagnosis can be made from the cytological, histological, and immunohistochemical findings. Local control of the disease can be achieved with complete and early resection of the tumor, and the benefit of adjuvant therapy is uncertain. Despite the rarity of this neoplasia, it should be part of the differential diagnosis when hemorrhagic cytological smears are obtained, especially when thyroid nodules causing compressive symptoms are present.

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