Hemangiopericytoma is a rare soft tissue tumor that may arise in many body sites. It is primarily a tumor of adult life and occurs in both sexes with equal frequency.

We report a case of thyroid hemangiopericytoma in a 58-year-old woman who presented with goiter known for 30 years. The tumor, 7x6 cm. in dimensions was located in the right lobe. Fine Needle Aspiration Biopsy (FNAB) with thyroid ultrasonography failed to be diagnostic and an open biopsy was made from the mass. Microscopic examination revealed malignant hemangiopericytoma. The case was accepted as inoperable. We present an uncommon tumor involving the thyroid with its clinical and histopathological properties in the light of the current literature.

Key words: Hemangiopericytoma, thyroid neoplasms

Introduction

Hemangiopericytoma (HPC) is primarily a tumor of adult life with a median age of 45 years. The tumor occurs in both sexes with equal frequency. Incidence among soft tissue sarcomas is 2.5% (1). Hemangiopericytomas may arise in many body sites (1, 2). The tumor is most common in the lower extremity, especially the thigh, pelvic fossa, and retroperitoneum. Less frequently it affects the abdomen and upper extremity. Location in the thyroid is exceedingly rare. Here we present a case of primary thyroid HPC with its clinical and histopathological properties.

Case Report

A 58-year-old women with goiter known for 30 years was admitted to the clinic with hardening of the thyroid gland. Thyroid ultrasonography demonstrated a lobular mass 7x6 cm in dimensions located in the right lobe. The mass contained calcifications and tiny cystic spaces. FNAB was made and a diagnosis of cystic colloidal goiter was rendered. Cervical CT revealed a mass lesion of malignant nature originating from the right lobe, infiltrating the isthmus, extending into the thoracic cavity, the subcutaneous fat, prevertebral fascia and destroying the hyoid bone. The larynx was displaced to the left. The mass had a heterogenous density and enhanced the contrast media. A frozen section was studied and diagnosed as malignant, so the case was accepted as inoperable. The sample of the tumor which was sent for frozen section was 2.5x1.5x0.8 cm. in dimensions and it was gray-white in color. Microscopically the tumor consisted of tightly packed cells around ramifying thin-walled endothelium-lined sacular channels ranging from small capillary-sized vessels to large gaping sinusoidal spaces. The cells had round to oval nuclei and moderate amounts of cytoplasm with ill-defined borders (Figure 1). The tumor was rich in cellularity and contained necrosis. Five atypical mitotic figures were observed in 10 High Power Fields. Tumoral cells were negative for Vimentin, Cytokeratin, S-100, F-8 LCA and SMA. Reticulin
preparation revealed a dense reticulin meshwork surrounding vessels and individual tumor cells (Figure 2). The tumor was diagnosed as malignant hemangiopericytoma.

**Figure 1.** A relatively uniform population of mesenchymal tumor cells intimately arranged about a prominent vascular network (HEx100)

**Figure 2.**

**Discussion**

HPCs comprise a group of benign and malignant soft-tissue tumors that may arise in almost all body sites. One fourth to one third of all HPCs arise in the lower extremities, especially the thigh. Approximately 10% to 25% are primary retroperitoneal tumors. An additional 10% to 16% arise in the head and neck region (3). HPC is primarily a tumor of adult life and has no sex predilection. Most patients with this tumor complain of a slowly enlarging painless mass. HPC of the thyroid is exceedingly rare (3-6).

HPC is composed of a relatively uniform population of mesenchymal tumor cells intimately arranged about a prominent vascular network, which includes irregularly shaped, dilated sinusoidal vessels. The inability to render an accurate assessment of the tumor’s biologic potential is the major problem for the pathologist. There are no prognostic criteria universally agreed upon. Some authors define a borderline category for HPCs. Among the features that contribute to borderline or malignant diagnoses are the presence of moderate cellular pleomorphism, more than slight mitotic activity, tumor necrosis, and a high level of cellularity. Sometimes vascular lumina are so compressed that they are difficult to recognize (1-3). The tumor cell configuration is ovoid or plump, rather than spindle shaped in borderline and malignant cases. Our case was considered malignant with its high cellularity, prominent mitotic activity and necrosis.

The HPC is thought to originate from the pericytes which are unique cells found spiralling around the outside of blood capillaries and post-capillary venules (2). Long processes extending from these cells and wrapping around the vessels can be demonstrated by silver staining techniques. The exact function of the pericyte is not known, but it has been postulated that this cell has contractile powers and so controls the caliber of the vessel.

It is sometimes difficult to predict clinical outcome with the histopathological features of HPC. Dictor and colleagues reported a thyroid HPC that was resected in a 5-year-old boy recurring in the larynx 8 years later (5).

As yet the application of FNAB in the primary diagnosis of soft-tissue lesions has only limited utility. It is very difficult to diagnose HPC with FNAB. Geisinger and his colleagues examined FNABs of three malignant HPCs from the head and neck region and one from the retroperitoneum and they compared their experiences with the histopathologic experience. They also studied the cases ultrastructurally and flow cytometric analyses were made. They concluded that, although a specific primary diagnosis of HPC on FNAB of a soft-tissue mass is unlikely, cytologic analysis might allow diagnosis of recurrent or metastatic HPC (3).

HPC is a very rare tumor of the thyroid and can invade neighbouring tissues. Primary diagnosis
with FNAB is extremely difficult. It has a characteristic and easily diagnosed histopathologic appearance. Both recurrence and invasion properties of the tumor make the prognosis hard to predict.

References


