Intravascular papillary endothelial hyperplasia (IPEH) is a proliferation of vascular endothelial cells that typically presents as a subcutaneous or submucosal mass. This entity was first described by Pierre Masson in 1923 as a hemangioendotheliovegevant intravasculaire and is perhaps better known by its eponym, Masson tumor. IPEH shares many clinical and histopathologic features with angiosarcoma so it is important to distinguish this benign proliferative process from malignant disease. IPEH can arise in any blood vessel; the presentation varies by location and can be quite heterogeneous. This case represents a rare clinical presentation of this entity, as a lateral neck mass. The treatment of IPEH is with surgical excision. Histology, radiology and the typical clinical characteristics of the diagnosis are discussed.

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) is a proliferation of vascular endothelial cells that typically presents as a subcutaneous mass. In the aerodigestive tract it may also present as a tender submucosal nodule or a bluish, raised mucosal lesion. The mass manifested in IPEH represents an exaggerated growth response by vascular endothelial cells as they respond to an inciting stimulus. It may present in a normal blood vessel, a vascular malformation, or as a response to local trauma. IPEH is generally believed to occur in the setting of a vascular malformation, or as a response to local trauma. IPEH is generally believed to occur in the setting of an intravascular location and mixed soft tissue density. The presence of calcifications is typical and the vessels involved may be occluded or patent. IPEH is composed of papillary processes covered by a single layer of endothelial cells. IPEH can arise in any blood vessel and the presentation varies by location and can be quite heterogeneous. Wang et al recently described a case of sinonasal IPEH that presented as chronic unilateral nasal obstruction. Another publication by Kim et al describes a case of recurring, unilateral submandibular swelling. In this case the lesion of IPEH was closely involved with the right submandibular gland and wharton’s duct, and was found to cause recurrent episodes of obstructive sialadenitis. Rare incidents of intracranial IPEH have been reported as well. Most recently Zhang et al published a case of IPEH of the jugular bulb and petrous temporal bone. In the current case, IPEH presented as a slow growing lateral neck mass. There are a limited number of similar reports in the literature. Based on the patient’s presentation our initial differential diagnosis included malignant disease such as squamous cell carcinoma, angiosarcoma, and lymphoma as well as more common benign lesions such as traumatic hematoma, vascular malformation, or neuroma. In summary, IPEH is a rare cause of a slowly growing neck mass. This condition is not a true neoplasm but rather reactive hyperplasia of the vascular endothelium. The appearance of IPEH on imaging studies can be quite variable and diagnosis is ultimately based on micropathology. Although intravascular procedures and radiation therapy have been studied, the primary curative management of these lesions is surgical excision.

Case Report

A 72-year-old female was seen in consultation with regards to a right neck mass. This mass was located in the right lateral neck. It had first been noticed 4 months earlier and had been slowly enlarging since. On exam the mass was noted to be superficial, freely mobile and nontender. The patient had no prior history of neck trauma or surgery.

A CT scan of the neck with contrast demonstrated a vascular mass (1.8 x 1.7 x 1.2 cm) just superficial to the sternocleidomastoid in the right neck. A fine-needle aspiration was performed which yielded grossly bloody fluid that was non-diagnostic. Subsequently, the mass was excised. Intraproactively, it was found to be vascular in nature and clearly arising from the external jugular vein. Pathologic evaluation of the surgical specimen ultimately revealed the diagnosis of IPEH.

Figure 1

Figure 2

The presentation and diagnostic evaluation demonstrated in this case is typical of IPEH. On CT the lesion was noted to enhance significantly with contrast and created a mass effect on the adjacent external jugular vein. It was noted to be heterogeneous in appearance with rim-like calcification (Figure 2). IPEH can have varied appearance on imaging studies depending on the degree of thrombosis and recanalization present. Important findings in this case include a likely intravascular location and mixed soft tissue density. The presence of calcifications is typical and the vessels involved may be occluded or patent. IPEH is composed of papillary processes covered by a single layer of endothelial cells. Microscopically the disease can be difficult to distinguish from angiosarcoma. Important distinguishing features have been described previously and include intravascular location, lack of nuclear atypia and mitoses, lack of necrosis, and lack of invasion of surrounding tissues. IPEH is a rarely encountered pathologic entity, however it most commonly presents in the subcutaneous tissues of the head and neck. The most common sites are generally regarded to be the scalp and face, followed by the oral mucosa. There are recent reports in the oral surgery literature of IPEH presenting as bluish mucosal lesions of the oral cavity. IPEH can arise in any blood vessel and the presentation varies by location and can be quite heterogeneous. Wang et al recently described a case of sinonasal IPEH that presented as chronic unilateral nasal obstruction. Another publication by Kim et al describes a case of recurring, unilateral submandibular swelling. In this case the lesion of IPEH was closely involved with the right submandibular gland and wharton’s duct, and was found to cause recurrent episodes of obstructive sialadenitis. Rare incidents of intracranial IPEH have been reported as well. Most recently Zhang et al published a case of IPEH of the jugular bulb and petrous temporal bone. In the current case, IPEH presented as a slow growing lateral neck mass. There are a limited number of similar reports in the literature. Based on the patient’s presentation our initial differential diagnosis included malignant disease such as squamous cell carcinoma, angiosarcoma, and lymphoma as well as more common benign lesions such as traumatic hematoma, vascular malformation, or neuroma. In summary, IPEH is a rare cause of a slowly growing neck mass. This condition is not a true neoplasm but rather reactive hyperplasia of the vascular endothelium. The appearance of IPEH on imaging studies can be quite variable and diagnosis is ultimately based on micropathology. Although intravascular procedures and radiation therapy have been studied, the primary curative management of these lesions is surgical excision. 

Discussion