Case Study: Atypical Presentation of Orbital Hemangiopericytoma

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ABSTRACT

Hemangiopericytoma is a slow growing, vascular mesenchymal tumor, which may behave like a malignant tumor, leading to local recurrence, or metastasis, or both. This report is about a 32-year-old lady with non-axial proptosis and diminished vision due to a mass lesion in the superomedial quadrant of the right orbit. CT scan and MRI reports showed no intracranial extension. Surgical intervention was done through the roof of the orbit keeping hemangioma, neuroma or schwannoma as the leading differential diagnoses; however, on aspiration, it was not bleeding. It was confirmed to be a hemangiopericytoma on histopathological examination. The unique presentation, where, on tapping, no blood was found, makes this case an example of diagnostic dilemma. This lack of bleeding might be due to vascular insufficiency of the mass. Proptosis disappeared the following day after the surgery and vision also showed significant improvement to 20/20 within a week after surgery. The patient has been kept on regular follow up keeping in mind the recurrent and metastatic behavior of the tumor.

Keywords: Bcl-2, CD-34, External Beam Radiation Therapy (EBRT), Pericytes, Proptosis, Pseudotumor, Spindle Cell Tumor, Stag Horn Vessels

INTRODUCTION

Spindle cell tumors are mesenchymal tumors having mesodermal origin. A hemangiopericytoma arises from pericytes. Another view is that it originates from pluripotent perivascular cells. In 1942, Stout and Murray described tumors, which were composed of capillary blood vessels with one or more layers of rounded cells arranged about them. These differed from capillary hemangioma because of the presence of perivascular cells and they named it hemangiopericytoma based on the morphological similarities with pericytes (Stout...
Pericytes were first described by Zimmermann in 1923.

Hemangiopericytoma is an unusual tumor representing only about 1% of all vascular neoplasms. It occurs in both genders with equal frequency in the head and neck region but is seen less frequently in the orbital region. It is more commonly found in the nasal cavity. It is responsible for about 1-3% of orbital lesions.

Dorfman first reported solitary fibrous tumours in 1994. After 1994 more cases of solitary fibrous tumors were detected. These were earlier labeled as hemangiopericytoma or hemangiopericytoma like tumors. Furusato et al (2011) published a study of 41 cases designated as hemangiopericytoma, fibrous histiocytoma and giant angiofibroma. They found that these tumors have overlapping morphologic and histochemical features and suggested solitary fibrous tumor be used as an all-encompassing terminology. According to Nappi et al (2012), a monomorphic population of compact polygonal or bluntly fusiform cells and stag horn stromal vascular pattern classically characterize a hemangiopericytoma. On the other hand solitary fibrous tumors are composed of variable pleomorphic spindle cells, admixed with collagen and arranged haphazardly in a patternless manner (Gengler C, 2006).

The distinction between solitary fibrous tumors and hemangiopericytoma can be subtle.

According to Gengler C et al (2006), the ramifying vascular pattern is not specific for hemangiopericytoma and can be seen in a variety of soft tissue tumors, like solitary fibrous tumor, fibrous histiocytoma, and other variants of solitary fibrous tumors. He no longer considered hemangiopericytoma a specific entity but rather as a growth pattern. He reclassified them as fibrous variant, giant cell variant, cellular variant, fat forming variant, and fibrous histiocytoma like variant.

Despite this resemblance, there are individual clinical features and distinctions on histochemical analysis. According to Westra WH et al (1994), a solitary fibrous tumor shows strong and consistent reactivity for CD34, with ropy collagen fibrils in contrast to a hemangiopericytoma, which hardly shows any focal reactivity to CD34 (Goldsmith et al 2001). Both these tumors show negative reactivity for immunodeterminants of epithelial and neuromyogenic differentiation (Keratin, cytokeratin SMA, S100). GenglerC et al (2006) reports reactivity against CD99 and Bel-2 could be positive in both hemangiopericytoma and solitary fibrous tumors.

Radiologically, in general, hypointense signals on T1W images and isointense to hypointense signals on T2W images indicates more of collagen content in solitary fibrous tumor. According to H J Kim et al (2008) hyperintense signals in T2W images could be due to cystic degeneration, hemorrhage or recent fibrosis. A hemangiopericytoma gives isointense signals on both T1W and T2W images. Hence, it is difficult to differentiate hemangiopericytoma from solitary fibrous tumors on MRI but it gives indication about collagen content, fluid content or hemorrhage within the tumor.

So, in a nutshell, according to available evidence, (Demmico EG et al, 2012) although hemangiopericytoma was once considered to be a distinct entity separate from solitary fibrous tumors, it is now considered to represent a cellular variant within the spectrum of solitary fibrous tumors. The term hemangiopericytoma refers to a particular morphologic pattern.
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