

Meningial Hemangiopericytoma

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Summary

Hemangiopericytoma is a potentially malignant vascular tumor that can occur anywhere in body, the meningeal location is rare. We report the case of meningeal hemangiopericytoma in 59-year-old man. The histological approach requires the use of immunohistochemistry (CD34, Vimentin, CD31, EMA, SMA, PS100, CD99, and Ki67) because this tumor is often confused with fibrous meningioma and solitary fibrous tumors. Thus, it is very important and crucial for the pathologist to identify the true nature of the tumor from the start, in order to make an appropriate prognosis.

The aim of this study was to report on the clinical, radiological and histological characteristics of meningeal hemangiopericytoma. A long follow up of these patients required because of frequent recurrences and delayed metastases.

Keywords: Hemangiopericytoma; Immunohistochemistry; Meningioma

Introduction

Primary meningeal hemangiopericytomas (MHPC) are uncommon accounting for less than 1% of primary Central Nervous System Tumors (CNST) [1]. This tumor arises from pericapillary cells or pericytes of Zimmerman [2-6]. MHPC formerly regarded as variant of angioblastic meningioma by Cushing & Eisenhardt [2,3,7,8]. In 1942, Stout and Murray identified a soft-tissue lesion with pericytes proliferation and called it a HPC [2,4,8]. The first documented case of primary intracranial HPC was reported by Begg & Garret in 1954 [3,8]. Despite all these evidences it is only in 1993 that MHPCs were classified as a distinct entity by the WHO [2,3,5]. Today, most pathologists are convinced that MHPC and soft-tissue HPC are similar. We report a case of MHPC specifying the contribution of immunohistochemistry in diagnosis of this tumour.

Observation

A fifty nine year old man was referred to our hospital with headache, dysarthria and the weakness in the left side of body of 2 months duration. The symptoms worsened by the onset of right hemiplegia.

A brain CT scan and MRI demonstrated a well-defined extra-axial lesion homogeneously and intensely enhancing 81/47/78 mm in the left posterior parieto-occipital area (Figure 1). The lesion was iso-intense on T1-weighted and T2-weighted with a central necrosis and perilesional edema. The lesion showed radiological imaging features suggestive of a meningioma. Total resection was performed.

Microscopic examination revealed perivascular proliferation of high cellularity composed of oval to slightly spindle cells with small cytoplasm and a large nuclear cytoplasmic ratio, in a densely hyalinised background and interrupted by numerous slit-like vascular spaces lined by flattened endothelial cells, so called "staghorn" sinusoids. Mitotic figures were present with two mitoses per 10 high-power as well as focus of necrosis. (figure.2). Immunohistochemical study (figure.3) showed positive reactivity weak and focal staining with CD34 strongly positive for vimentin and CD99; but negative for epithelial markers such as (EMA, keratin), PS100 and Bcl-2. Ki67 staining index was zero. The histological and immunohistochemical features confirmed the diagnosis of HPC.

The patient received post operative radiation therapy.

Discussion

MHPC are rare, aggressive neoplasms that are mainly located in the musculoskeletal system and skin [3,7]. MHPC constitutes approximately 0.4% of all primary CNS tumors [2] and 2% to 4% of all meningeal tumors [3,5].

Unlike meningiomas MHPC tend to occur more often in man than women with a M/F ratio 1.4/1 [2]. The average age of diagnosis is in the fourth [2,9] and fifth decades [3].

MHPCs present a similar distribution to those of meningiomas, essentially supratentorial in 77% in the series reported by Ksira et al. [5] as in our case.

The clinic depends on the location [2,9] and size of the tumor, when the seat is supratentorial, headaches are the main symptoms, and seizures (15-30%), a sensory or a motor deficit (40%) [9].

Imaging features, MHPC are difficult to differentiate from meningiomas and Solitary Fibrous Tumours (SFT) [3]. On CT scan MHPC show a sharply demarcated tumor with a broad-based dural attachment [2,7].

On MRI these tumors are isointense to cortical gray matter on T1WI and isointense to slight hyper-intense on T2WI [2,3,7,9] post-injection films demonstrate a intense enhancing extra-axial lesions [3]. Our observation was similar to radiological findings.

Angiography has showed an anarchic hyper-vascularisation with a marked blush tumor [5,9,10]. In macroscopy, MHPC is a globoid slightly lobulated tumor, well-demarcated from the adjacent brain tissue and is tendency to bleed during removal, sometimes infiltrated [2].

Microscopic examination reveals highly cellular monotonous tumors composed of round or fusiform cells with scant cytoplasm, oval

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nuclei, small nucleoli and they lack the pseudo-inclusions [2,3,4]. A rich reticulin network supporting prominent “Staghorn sinusoids” is commonly observed. Frank necrosis is uncommon and calcification is not a usual feature [11].

Immunostaining for MHPC show an intense reactivity to vimentin but not to EMA, unlike meningioma that is positive for both [2,3,12]. CD34 appear patchy or weakly staining in the HPC [3] as in this case but diffusely staining in the SFT. Bcl-2 seems to be discriminate of differentiate these two entities. In fact, SFT is usually intensely positive to Bcl-2 when MHPC is negative to this body [4]. CD99 is thought to be a good marker for HPC with specificity about 84% as in our study [4].

Besides, MHPC also shows a negative reaction to S-100 protein, classical endothelial antigens (factor VIII, CD31) as well as progesterone receptor [2]. Focal positivity to desmin, smooth muscle actin and cytokeratin may be occasionally encountered [2,12,13]. Labelling indices for Ki67 ranged from 0.2 to 39 % [12]. It was zero in our result.

Electron microscopy demonstrates the presence of small bundles of intermediate filaments surrounded by basal lamina-like amorphous material characteristic of HPC [2,3,13].

Cytogenetic studies have reported rearrangements of chromosome 12q13 and alterations on 6p21, 7p15 and 19q13 [2,3,13].

These tumors are treated by total resection followed by radiotherapy [2,3,5,6].

The biological behaviour of MHPC is usually malignant and tends to recur in many years. Therefore long- term follow-up with annual serial imaging is recommended [3,12].

Conclusion

We have reported a case of MHPC and a review of literature. MHPC is a well-defined clinicopathologic entity exhibiting high rates of recurrences and late extracranial metastasis, for this reason a long-term clinical and radiographic follow-up is mandatory for these tumors.

It must be distinguished from fibrous meningiomas and SFT. The histological diagnostic certainty is complemented by

immunohistochemical and ultrastructural studies. Surgery followed by radiotherapy is the mainstay of the treatment.

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