Case Report

Recurrent Intracranial Haemangiopericytoma compatible to Synovial Sarcoma

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Abstract

A rare case of intracranial haemangiopericytoma with a thrice recurrence, treated by gross total removal and local irradiation is presented. Histological examination of the tumour specimen showed haemangiopericytoma (WHO grading III). The tumour has not recurred for 15 months after third operation and 30 sessions of radiotherapy, although the effectiveness of radiation for haemangiopericytoma is unclear.

Introduction

Haemangiopericytoma (HPC) was first described and named by Stout and Murray in 1942, as a rare vascular tumour originating from Zimmermann's pericytes around capillary and post capillary venules. Intracranial Haemangiopericytoma is another rare, highly vascular tumour accounting for less than 1% of all tumours of central nervous system (CNS) or accounts for 2-4% of meningeal tumours. This neoplasm is indistinguishable from haemangiopericytoma at other sites and is no longer considered variants of meningioma, because genetic analysis has shown no relationship between haemangiopericytoma and meningiomas.[1] The current classification of World Health Organization (WHO classification 1997 and 2000) introduced this neoplasm as a specific tumour of CNS and as an entity by its own.[2] HPC is considered to be malignant, recurs in 50%-80% of cases in series of 20 or more patients and metastasizes in approximately 23.4% cases.[1] In the present study we describe a Haemangiopericytoma and its recurrence thrice, which was initially diagnosed as synovial sarcoma.

Case Report

A 27-years-old man was admitted to Imam Hussein hospital, Kermanshah University of Medical Sciences, Iran in September 2003 with the complaints of fatigue, nausea,

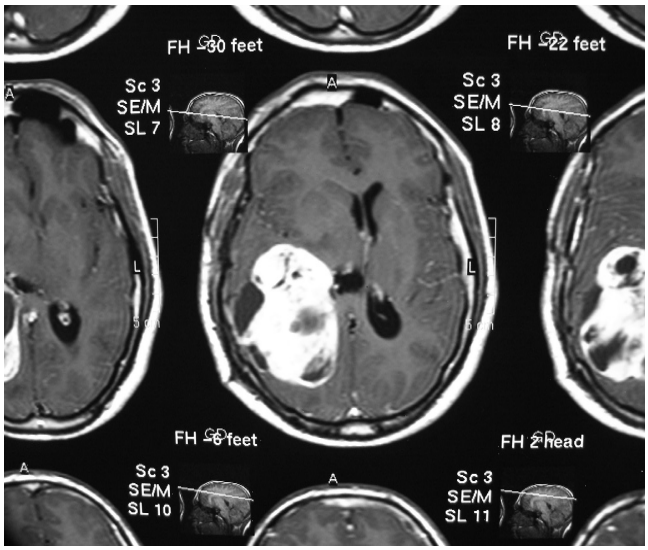

Figure-1: Horizontal section, T1-weighted MRI before his second operation.

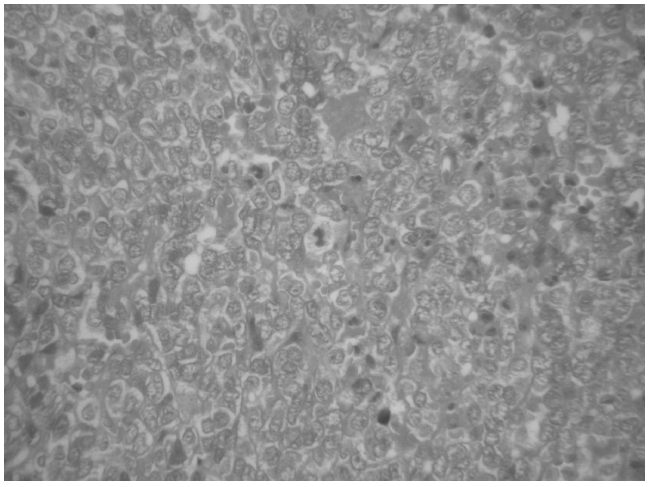

Figure-2: High vascular tissue and polymorphism in H&E staining section ×100.

headache, blurred vision and disequilibrium for the last one year. Magnetic Resonance Imaging (MRI) of the brain revealed a large enhanced lesion in left parietooccipital lobe (Figure-1). The tumour was removed surgically. Histological study confirmed it to be an "angioblastic menangioma". Two years later the tumour relapsed in parietooccipital and posterior lobes. Surgery was performed again and histological study showed "malignant ependymoma" as a second diagnosis. In March 2006, one year after reoperation, the patient complained of headache and nausea. Tumour resection was performed again; the lesion was soft and bled easily on touch. After the third surgery, the patient underwent adjuvant radiotherapy for 30 sessions. Based on the morphological and immunohistochemical features of a synovial sarcoma, a lesion morphologically closely related to haemangiopericytoma, was diagnosed. More examinations were performed on paraffin embedded blocks. At this time, H&E stained sections showed a rather cellular, highly vascular tumour (Figure-2). It was composed of cells with ill defined borders and moderately polymorphic, round or elongated nuclei with sparse chromatin. No specific histoarchitecture was recognized, however some of the numerous thin-walled vascular spaces displayed a typical "staghorn" shape. In addition, geographic necrosis and mitotic figures were present.

In immunohistochemical staining, the tumour cells were immunoreactive for vimentin and focally (but widespread) also for CD99 and BCL2. CD34 staining highlighted the abundant vascularization, but also showed a very focal immunoreactivity of tumour cells. The tumour was not immunoreactive for epithelial membrane antigen (EMA), smooth muscle actin, desmin, pan-cytokeratin (AE1/AE3), synaptophysin, glial fibrillary acidic protein (GFAP) or myogenin. The proliferation rate (MIB-1 (Ki67) staining) was about 10%. Fluorescence in-situ hybridization (FISH) did not disclose a T(X, 18) translocation. Synovial sarcoma was considered in the differential diagnosis at that time, however absence of typical T(X, 18) translocation made this diagnosis unlikely. In fact, the morphology of the tumour and its immunohistochemical profile were typical for haemangiopericytoma. The considerable mitotic activity in this tumour, the presence of necrosis and high MIB1 labeling index justified its grading as WHO III.

Discussion

HPCs are rare mesenchymal tumours that may arise in the head and neck, trunk, skin, retroperitoneum and oral cavity. In the central nervous system, HPC is a highly cellular and vascular tumour which is indistinguishable histologically from similar lesions occurring in the peripheral soft tissues.[3] Haemangiopericytoma originating in the intracranial meninges was first reported by Beggs and Garret.[4] Some authors have classified these tumours as angioblastic meningioma, but others consider it to be distinct from meningioma and represent a type of vascular neoplasm. Aggressive growth, tendency to local recurrence and relatively frequent metastases are clinical features of the tumours.[3]

HPCs frequently recur and metastasize extracranially in contrast to ordinary meningiomas.[5] Goellner et al. reported local recurrence in 80% and extracranial metastasis in 23% of 26 patients with haemangiopericytomas.[6] Itoyama et al.[7] consider that HPCs should not be classified as meningiomas, because the electron microscopic appearance is very different to meningioma and the incidence of recurrence and

extracranial metastasis is much higher. The treatment of this tumour is based on the complete surgical resection. Post-operative radiotherapy is strongly recommended, even after an apparently complete tumour resection.[8] We consider the radiation therapy to have been effective in our case, as there has been no tumour recurrence for 15 months after the third operation.

Prediction of patient outcome is difficult based on current knowledge about the tumours and histological parameters only. However, on the basis of an analysis of central nervous system haemangiopericytomas, rapid regression is correlated with increased mitotic rate ($\geq$ 5 mitotic figures per 10 high-power fields), high cellularity, nuclear pleomorphism, haemorrhage and necrosis, in accordance with that of peripheral HPCs.[8]

In histopathological studies, HPCs have a particular profile, which aid in the differentiation of this tumour from other similar lesions such as meningioma or synovial sarcoma. The tumour cells react with antibodies against vimentin and in most cases, against CD34, but unlike meningioma, they lack epithelial membrane antigen (EMA) and also unlike the synovial sarcoma do not disclose T(X, 18) translocation.[8] In microscopy, a dense reticulin network typically investigating individual tumour cells is one of the most characteristic features of the tumour. The appearance of "staghorn-like" vessels separates the tumour cells into small lobules, the feature which presents in synovial sarcoma. Synovial sarcoma is a mesenchymal spindle cell tumour that displays variable epithelial differentiation including glandular formation.[9] In approximately 10-20% of all cases of synovial sarcoma, there is a distinctive haemangiopericytoma-like vascular pattern.[10] This pattern, however, does not display a broad range in the number of vascular channels that are present and is almost always associated with distinct spindle cells and sometimes myxoid or calcified areas.[9] To make a safe differential diagnosis between synovial sarcoma and haemangiopericytoma, many tumour sections should be examined and immunohistochemical staining is a necessity. In our patient the lesion appeared to be a synovial sarcoma at first and was later found to be a haemangiopericytoma (WHO grade III). The cytogenetic hallmark of synovial sarcoma is T(X, 18) (p11.2; q11.2), which was not present in our patient. In this case complete surgical removal and radiation therapy was opted as it is considered to be effective, and there was no tumour recurrence till 15 months after the last surgery.

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