A RARE CENTRAL NERVOUS SYSTEM TUMOR: GLIOSARCOMA, CASE REPORT

NADİR BİR SANTRAL SİNİR SİSTEMİ TÜMÖRÜ: GLİOSARKOMA, OLGU SUNUMU

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ÖZET

Gliosarkoma primer merkezi sinir sistemi tümörlerinin nadir bir formudur. 2007 Dünya Sağlık Örgütü (WHO) kriterlerine göre glioblastomanın bir varyantı olarak kabul edilir. Sıklıkla serebral hemisferleri tutar. Gliosarkomlar malign glial ve mezenkimal komponent içeren bifazik tümörlerdir. Gliosarkomun glioblastomdan ayırımında immunohistokimyanın önemi büyüktür. Gelişim mekanizmaları net değildir. Büyük kısmının de novo olarak geliştiği daha az bir kısmının glioblastome multiformenin daha önce bu sebeple rezeksiyon ve radyoterapi yapılmış alanlardan geliştiği de düşünülmektedir. Yapılan moleküler genetik çalışmalar gliosarkomun her iki komponentinin de monoklonal prekürsör hücrelerden köken aldığını öne sürmektedir. Çalışmamızda acil servise baş ağrısı ile başvuran 53 yaşında erkek hastadaki gliosarkom olgusu sunulmaktadır. Gliosarkomların klinik, histopatolojik ve immünhistokimyasal özelliklerini tanımlamak ve patogenetik mekanizmalırını tartışmak hedeflenmiştir.

Anahtar Kelimeler: gliosarkoma, gliobalastom, immunohistokimya

SUMMARY

Gliosarcoma is a rare tumor of the central nervous system composed of glial and mesenchymal components. It is considered to be a glioblastoma variant according to the 2007 World Health Oranization (WHO) classification and often holding the cerebral hemispheres. Gliosarcomas are biphasic tumors that containing malignant glial and mesenchymal components. Immunohistochemistry is important to distinguish gliosarcoma and glioblastoma. Developmental mechanisms of gliosarcomas are not clear. It is considered that large part of gliosarcomas develop as de novo, and lesser part of them develop after resection and radiotherapy for glioblastome multiforme. Molecular genetic studies suggest that the two components of gliosarcoma origin of monoclonal precursor cells. In this study a case of gliosarcoma in 53 years old man who applied to emergency room with headache, is reported. We aimed to describe clinical, histopatological and immunohistochemical features of gliosarcoma and to discuss its pathogenetic mechanisims.

Keywords: gliosarcoma, gliobastoma, immunohistochemstry

INTRODUCTION

Gliosarcoma a rare form of primary central nervous system tumors.[1,2] It is considered to be a variant of glioblastoma according to 2007 World Health Organization (WHO) criteria (Grade IV tumors).[3,4] Gliosarcomas include 2% of all glioblastoma patients, 5% of astrocytomas and 8% of anaplastic astrocytomas. Often they are detected between the ages of 40-60.[5] Gliosarcomas are more common in males (M: F = 1,8:1), and often holding the cerebral hemispheres that are seen in order of frequency temporal, frontal, parietal, occipital areas. Gliosarcoma described for the first time in 1895 by Stroebe. In 1955, Feige et al. was defined gliosarcoma as a variant of the glioblastoma which have sarcomatous areas with vascular proliferation.[6,7].

In our study, located in the right frontal intracranial tumor which was diagnosed as gliosarcoma according to morphological and immunohistochemical findings, are presented.

CASE REPORT

53-year-old male patient was admitted to our outpatient clinic complaining of a headache. With cranial magnetic resonance imaging (MRI) examination, intracranial mass was determined. Patients underwent right frontal craniotomy and gross total excision of the tumor tissue. There is no known medical history of the patient.

In gross examination; 15 cc volume of bleeding, creamy-white colored tissue fragments which had locally nodular areas was seen.

In light microscopy examination with hematoxylin-eosin (H & E) sections; observed locally spindle, hyperchromatic nuclei, eosinophilic cytoplasm cells with mitotic figures that mimicking herring bone pattern and meanwhile clusters of glial cells containing large eosinophilic cytoplasm, orthochromatic large nuclei and mitosis (Figure 1).

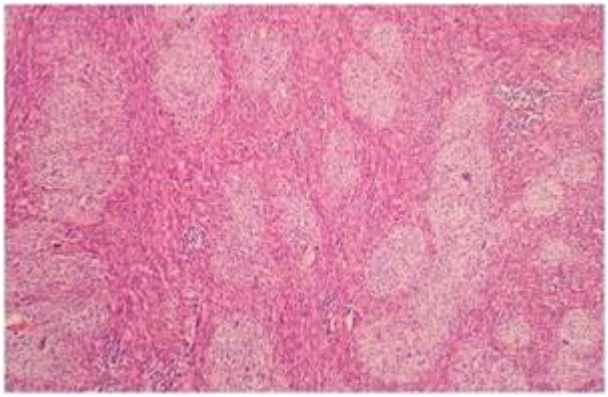

Figure 1 *Glial and sarcomatous areas (H&E, 40x)*

Immunohistochemically, there was a biphasic staining pattern for GFAP and vimentin. In mesenchymal areas there was a strong positive staining with vimentin and no staining with GFAP, however, in glial areas a strong staining for GFAP and no staining with vimentin was observed (Figure 2, 3). Ki 67 proliferation index was 30%, the number of mitotic figures was 32/10 HPF. The present findings are evaluated as a gliosarcoma.

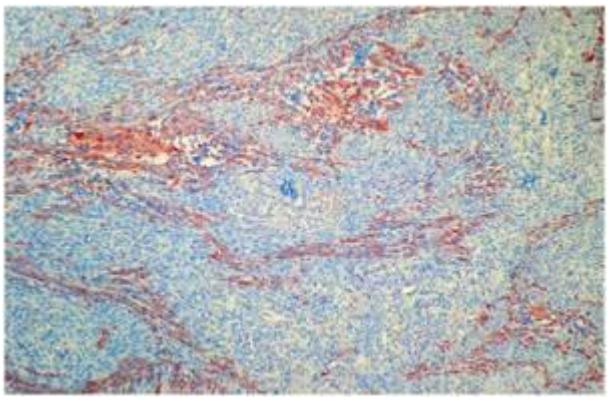

Figure 2 *Positive staining with GFAP in glial components (GFAP, 100x)*

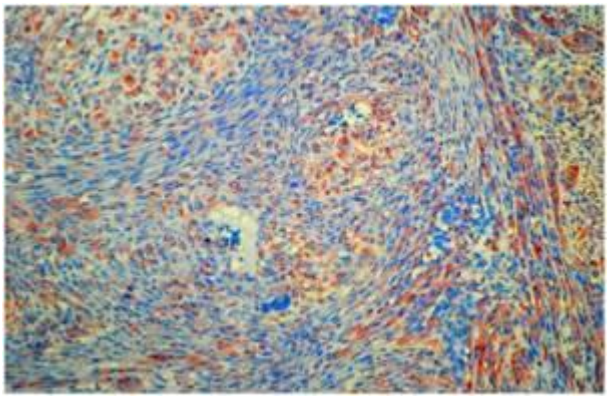

Figure 3 *Positive staining with vimentin in sarcomatous areas (vimentin, 200x)*

DISCUSSION

Gliosarcomas are biphasic tumors that containing malignant glial and mesenchymal components.[1,2,5] Glial component is a high-grade malignant astrocytic component that contain adenoid or squamous metaplasia areas.[8] Typically, the sarcomatous component is fibrosarcoma or malignant fibrous histiocytoma which may include neoplastic smooth muscle cells, endothelial cells, and more rarely, chondroid and osseous elements.[6]. In our case sarcomatous component resembled a fibrosarcoma in looks.

Immunohistochemistry is important to distinguish gliosarcoma and glioblastoma. Strong GFAP staining of glial component, absence or characteristic biphasic, very weak staining of sarcomatous component is very important for the diagnosis.[6] Our case showed a similar immunohistochemistical features to the literature.

Developmental mechanisms of gliosarcomas are not clear. It is considered that large part of gliosarcomas develop as de novo, and lesser part of them develop after resection and radiotherapy for glioblastome multiforme.[2] In transformation theory is one of the theories of the development mechanism of gliosarcoma, indicated that in a portion of the glial tumor, dedifferentiation of primitive cells results in sarcomatous component transformation. Because of this theory; considered that due to the applied radiation to pre-existing tumor of the central nervous system initiate dedifferentiation of glial cells to sarcomatous cells.[4] There was no history of resection or radiotherapy of our patient suggested that gliosarcoma developed as de-novo.

Molecular genetic studies suggest that the two components of gliosarcoma origin of monoclonal precursor cells.[9] Biernat et al. revealed that the monoclonal theory first. In their study; demonstrated

that P53 mutations in both components.[10] In some cases, both components of gliosarcoma the presence of p53 and p53 nuclear protein and immune revealed that the idea of both glial and sarcomatous structure composed of neoplastic glial cells.[1]

Histogenesis of gliosarcoma is controversial. Feigin and colleagues defined sarcomatous component, as the proliferation of blood vessels glioblastoma. However they could not be determined endothelial origin in sarcomatous component with immunohistochemical study.[11] In the study of Reis et al. which they investigate the genetic profile of gliosarcomas PTEN mutation, nuclear accumulation of P53, deletion of P16 and amplification of CDK4, was seen in both glial and sarcomatous components.[12]

Prognosis of gliosarcoma is very poor. Sarcomatous component is not important for prognosis, therefore it is similar to glioblastoma.[6]

Gliosarcomas have worse prognosis in patients with extracranial metastases. Extracranial propagation is effected via Cerebrospinal fluid (CSF), and local recurrence is common.[8] MIB-1 (Ki67) score is a grading system is used for soft tissue sarcomas, it is also a prognostic factor for glioblastoma and gliosarcomas, varies between 7.7% 36.1%.[7] In our case, the MIB-1 index was found 30%. The patient died 6 months after diagnosis.

Consequently; gliosarcomas are rare tumors of central nervous system. Histopathological examination and immunohistochemical studies' role is important in the diagnosis. Most of them accept that devoloped as de novo.

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