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EDİTÖRE MEKTUP / LETTER TO THE EDITOR

Malignant glioma arising from orbital teratoma in childhood

Çocukluk çağında orbital teratom zemininde gelişen malign gliom

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To the Editor,

Teratomas contain components of three germ layers (endoderm, mesoderm and ectoderm) and usually found in the gonadal regions, pineal gland and mediastinum. Orbita is an uncommon location for teratoma and only individual case reports can be cited in medical literature[1,2]. Here, a pediatric patient with an orbital tumor diagnosed malignant glioma arising from malignant teratoma is reported.

An 18-month-old female was referred to our center because of a diagnosis of optic glioblastoma (malignant glioma) after enucleation of the left eye. In patient history, it was leant that the mass in the left orbita was present in neonatal period. The patient was brought to another hospital when she was 6-month-old. Tru-cut biopsy from the orbital mass revealed no definitive diagnosis and enucleation was conducted. Pathological examination revealed malignant teratoma with a malignant glioma (glioblastoma) component. It was thought that malignant glioma arised from malignant transformation of an orbital teratoma because of intraorbital localization of the tumor, choristomatous elements including abnormal and neoplastic epithelial structures and disorganized ocular tissue not consistent with their anticipated anatomic location. The malignant component was a spindle cell tumor with vascular proliferation, necrosis and lots of mitotic figures compatible with malignant glioma (glioblastoma). No involvement of the tumor to the optic nevre or extension from optic tract was reported. Despite tratment with bleomycin, etoposid and cisplatin the tumor relapsed and second line treatment consisting of vincristin, doxorubicin and cyclophosphamide was given. Chemotherapy was ceased when the patient was 24-month-old. The patient is well and continues her controls at outpatient clinic for 14 months.

Orbital teratomas are rarely malignant but usually present as benign and localized mass. Globe is usually not affected but compressed and pushed forward resulting in marked proptosis[2]. Some patients with an orbital teratoma have intracranial or periorbital involvement or the tumor may originate and extend from these sites. In our patient, orbital tumor was present at birth but no intracranial or periorbital extension was shown neither in radiolagical images nor in pathological examination.

Clinical or radiological differentiation of orbital teratomas from other benign or malignant neoplasms is difficult. Computerized tomography and magnetic resonance imaging can provide clues about the presumptive diagnosis in some patients. However it is best to undertake an orbital biopsy, either excisional or incisional if the lesion is in a surgically accessible location. Fine needle aspiration biopsy can not always reveal correct diagnosis and surgical excision of the mass is recommended for definitive diagnosis[3]. Although some authors suggest early surgery there are many reports in which the eye has been saved. In our patient, since tru-cut biopsy did not reveal any definitive diagnosis and the tumor was massive and globe could not be clearly delineated an orbital enucleation was undertaken.

Although teratomas are commonly benign tumors

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their malignant potential may not be clear until they are excised completely. When the neural tissue is evident like fetal neural elements it is said to be immature or malignant[4]. The present case showed masses of undifferentiated cells resembling primitive neuroepithelium, indicating its malignant nature and there was no intracranial extension. The malignant component in our patient was a spindle cell tumor with vascular proliferation, and necrosis and numerous mitotic figures were compatible with malignant glioma. Malignant glioma was thought to be arise from malignant transformation of an orbital teratoma because of intraorbital localization of the tumor, choristomatous elements including abnormal and neoplastic epithelial structures and disorganized ocular tissue not consistent with their anticipated anatomic location.

Optic nevre gliomas are generally accepted as low grade astrocytomas. They generally ocur in children younger than 10 years of age[5]. Optic gliomas demonstrate an association with neurofibromatosis type 1. No intracranial or optic tract extension was shown neither in radiolagical images nor in pathological examination in the present case. Additionally, there was no finding in physical examination and in radiological images compatible with neurofibromatosis type 1. Recurrence after enucleation and first line chemotherapy is consistent with the aggressive nature of the malignant glioma.

Our case is unique because we could not find any similar case, reporting malignant glioma arising from an orbital teratoma in literature search.

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