EUR. J. ONCOL.; Vol. 23, n. 1, pp. 57-59, 2018

A case of ependymoma with unusual radiological presentation

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Summary. Ependymomas are glial tumors, usually seen as intra-axial lesions in MRI. Here we report a case of an extra-axial lesion, resected as a meningioma; but pathology report was a Myxopapillary Ependymoma. Further evaluation detected another lesion in distal part of spinal canal too. So it seems logic that extra-axial tumors be managed with more caution.

Key words: ependymoma, meningioma, craniospinal irradiation, extra-axial, MRI

Introduction

Ependymomas are glial tumors that arise from ependymal cells within the CNS. Patients with Ependymoma are usually pediatric cases and the location of their tumors isintracranial, while in adultsEpendymomas are usually located inspinal canal (1).

Majority of the intracranial Ependymomas are located in the posterior fossa (infratentorial) usually arising from the floor of the fourth ventricle (1-3) (60%), while the remainder are located supratentorially(40%) (4). Supratentorial Ependymomas that are extra-axial are very rare, with only a few reported cases in the literature.

The World Health Organization (WHO) divides Ependymomas into 4 types on the basis of histologic appearance (5):

- WHO grade I: Myxopapillary Ependymoma, Subependymoma
- WHO grade II: Ependymoma (with cellular, papillary and clear cell variants)
- WHO grade III: Anaplastic Ependymoma

Case report

In September 2016, a 24 year old Iraqi girl came to our center becauseof nausea, imbalance, vertigo, and also nervousness which have been progressing since 2 years ago. A brain MRI without Gadolinium was done for the patient. A huge lobulated extra axial brain lesion, embedded on left Sylvain fissure with extension to Paraseller region was seen in this MRI (Figure 1).

Radiological diagnosis for this lesion was Meningioma.Due to the large size of tumor (5.8*3*3.8 cm), patient was referred for surgical resection in neurosurgery department. The removed creamy-brown rubbery specimen got examined under microscopic evaluation. Surprisingly, the primary pathologic report was suggestive for myxopapillary epandymoma. So, confirmatory IHC study by checking CK, GFAP, S-100vimentin and EMA was done, and the diagnosis was confirmed!

Theneurosurgeon referred the patient to Radiation-Oncology department for further evaluation and also complementary treatment. An MRI with and without Gadolinium for the whole spine of the patient was done. An abnormal signal 14*13 mm intradural extramedullary hetergenous enhanced mass lesion at the L4-L5 level was detected (Figure 2). The neurosurgeon refused to do a surgical resection for this new lesion. So we decided to do radiation treatment. In the first phase, a craniospinal irradiation (CSI) was done up to the dose of 36Gy in 20 fractions. A boost dose to the surgical bed of the resected lesion in the brain,

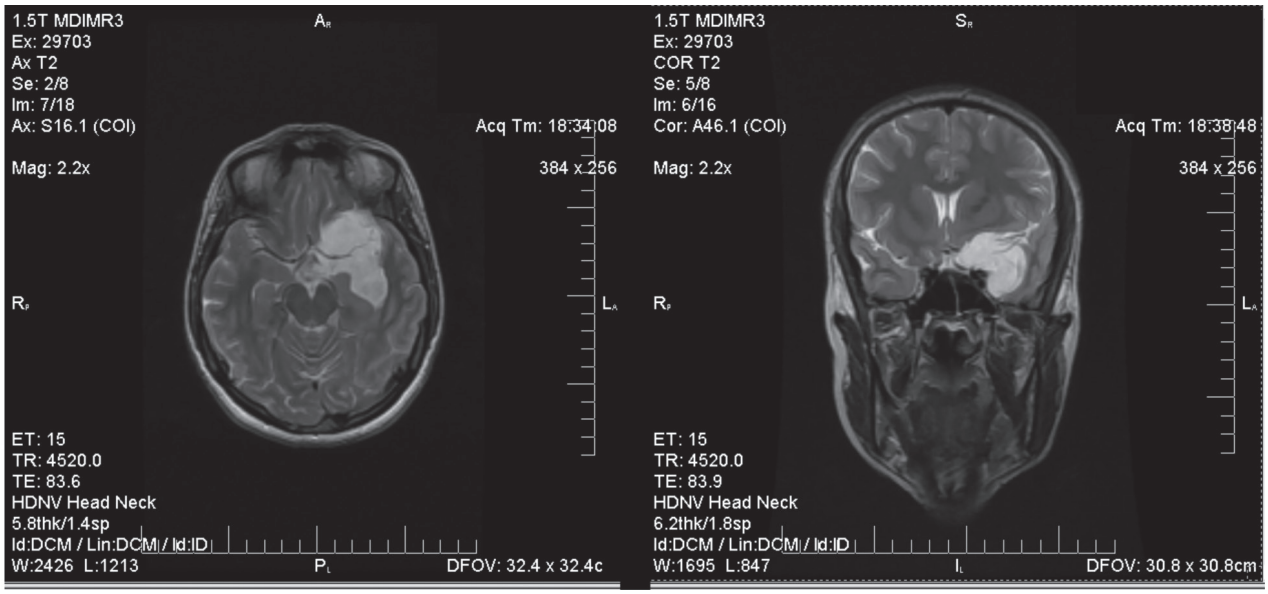

Figure 1. Pre-operative brain MRI (T2-FLAIR). A huge lobulated extra axial enhanced brain lesion, embedded on left Sylvain fissure with extension to Para seller region

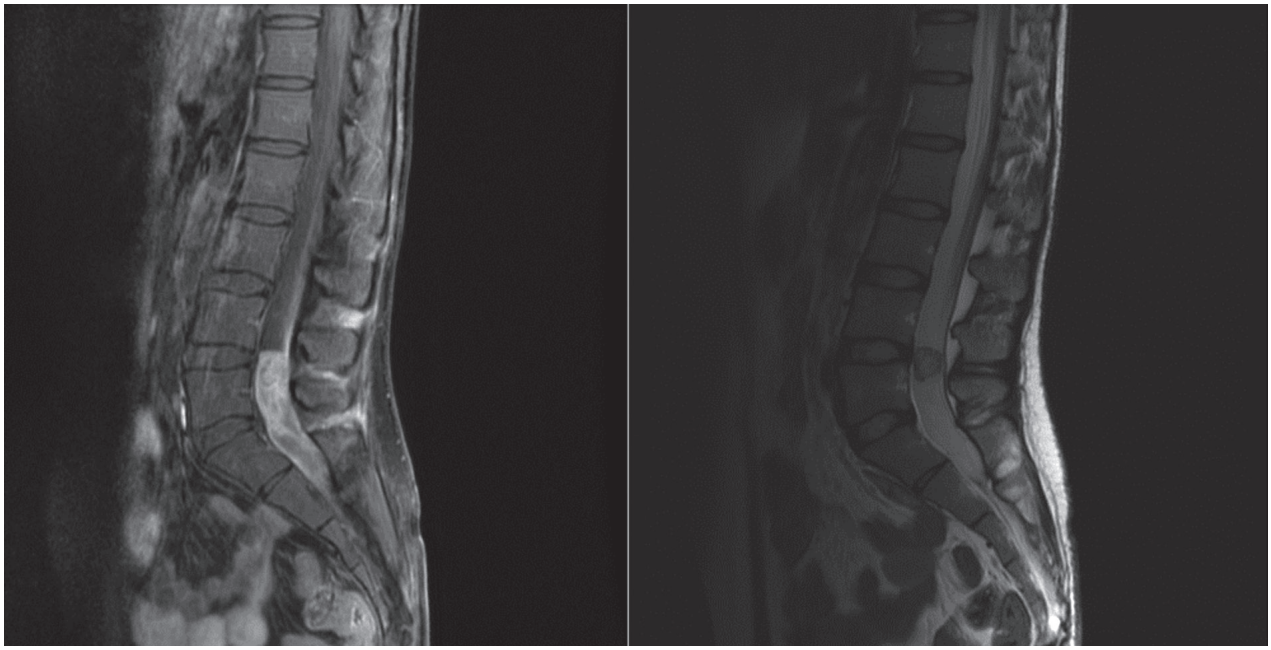

Figure 2. Intradural extramedullary hetergenous enhanced mass lesion at the L4-L5 level. (T1 with contrast [left] and T1 without contrast [right])

as well as to the lesion in the spinal canal was delivered up to the cumulative dose of 54Gy in 30 fractions. The patient tolerated the treatment very well. Control MRIs after treatment were done in Iraq. After about one year, the lesions are controlled without any progression.

Discussion

Ependymomas are usually intramedullary tumors. Extradural presentation with invasion of surrounding tissues is extremely rare (1).

Several hypotheses have been put forward to explain the origin of extra-axial Ependymomas with no connection to the ventricles. Fukui et al., proposed that such a tumor arises from glial rests in the subarachnoid space to produce an extra-axial mass (6). Hayashi et al., suggested that the tumor originates around the ventricle, grows and extends extramedullarly, followed by degeneration and necrosis of the ventricular portion of the tumor, leaving an extra-axial Ependymoma (7). According to Lyons et al., grossly nonvisible microscopic cellular tracts exist in development, between the ventricle and extra-axial Ependymoma that facilitate tumor extension into the subarachnoid space. These extensions subsequently regress (8). Vernet et al., postulated that tumors develop from intraparenchymal or subarachnoid ependymal cysts that result from disorders of migration from the germinal matrix (2, 6, 9). They represent primitive neuroectodermal tumors that have differentiated extensively along the ependymal lineage and might be the result of neoplastic growth within ectopic ependymal cells and are the consequence of a migration error (2, 10).

In conclusion, it should be taken into consideration that one of the differential diagnoses of the extra–axial lesions could be Epandymoma. Although there are no recommended guidelines for the management of extra-axial Ependymomas, it is advised that gross total resection should be done in such patients. Specially in children and young adults, it is more important when the patient is candidate for non-surgical treatment techniques like radiosurgery thereupon no pathologic evidence will be obtained. In these situations we should consider probable differential diagnoses like Epandymoma and if needed more evaluations like whole axis imaging should be performed (11).

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