CASE REPORT

Adult Desmoplastic Medulloblastoma

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A B S T R A C T

Medulloblastoma is more common in young age and rare in adult age. Some characteristics that characterize medulloblastoma in adults compared with children: Lateral cerebellar location, heterogeneous signal intensity on magnetic resonance imaging, desmoplastic histological variant and more favorable prognosis. Preoperative diagnosis is important for correct management of these patients. However, because of the low incidence of medulloblastoma in adults, preoperative diagnosis still challenging and prognostic factors and best treatment options are still controversial. We report a case of a 31-year-old male patient who presented with a rare case of posterior fossa medulloblastoma.

Key words: Adult, cerebellar, desmoplastic, medulloblastoma

ملخص البحث:
يكثر الورم النخاعي (medulloblastoma) لدى صغار السن ويندر لدى البالغين. يتميز هذا الورم لدى الكبار مقارنة بالصغار بما يلي: موقع المخيخ الجانبي، الإشارات غير المتجانسة في الرنين المغنطيسي، البديل النسيجي الصلد والنتائج الجيدة. التشخيص قبل إجراء الجراحة ضروري لعلاج هؤلاء المرضى ولكن لندرة حدوث الورم لدى البالغين فإن التشخيص قبل إجراء الجراحة يظل من التحديات وتظل بدائل العلاج أمراً مختلف عليه. يعرض الباحثون حالة لمريض في الحادية والثلاثين من العمر كان يعاني من هذا الورم في التجوف الخلفي.

INTRODUCTION

Medulloblastoma is a primitive neuroectodermal tumor (PNET), and it is the most frequently encountered primary brain tumor in children. About 50% of these tumors occur in children aged <5 years, whereas they are less common in adolescents and young adults. Medulloblastoma's behavior is different in adults than in children, and it is identified as a different biological and clinical entity.[1]

Adult medulloblastoma has a higher proportion of desmoplastic histological characteristics than medulloblastoma in children, and it has a higher incidence within the cerebellar hemispheres, thus featuring different proliferative and apoptotic indices and having a tendency for late relapse.[2]

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CASE REPORT

A 31-year-old, Saudi soldier living in Hafr Al-Batin presented with a headache, vomiting, blurring of vision, positive Romberg test and reported a gait disturbance of 1-month duration. The patient was full power, no cranial nerve deficit and Glasgow coma score 15\15.

A computed tomography (CT) of the brain showed a right cerebellar heterogeneous mass a ffecting the fourth ventricle with dilation of the third and lateral ventricles with transependymal edema. It was associated with crowning at the foramen magnum denoting impending herniation [Figure 1a and b].

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Emergency external ventricular drain (EVD) was inserted. After EVD, the patient improved [Figure 1c]

A magnetic resonance imaging (MRI) showed a right-sided cerebellar mass measuring 4 cm × 3.5 cm × 3.8 cm, with heterogeneous signal intensity. This lesion had an anterior cystic nonenhancing component and a posterior enhancing solid component. The lesion had a significant mass effect with attenuation of the fourth ventricle [Figure 2].

A few days later, the patient underwent elective surgery, a suboccipital craniotomy with gross total mass excision, which was confirmed by a postoperative follow-up MRI [Figure 3].

Ten days after surgery, the patient had suboccipital wound site fullness and cerebrospinal fluid (CSF) leak, which required the insertion of a V-P shunt.

Histologic examination of the mass showed a markedly cellular neoplasm composed of rounded, oval or elongated, primitive cells arranged in reticulin-free nodules encircled by reticulin-rich, more densely packed zones [Figure 4a and b]. Such a mass is known as a desmoplastic because it is permeated by fine collagen (reticulin) fibers that give it a firm consistency.

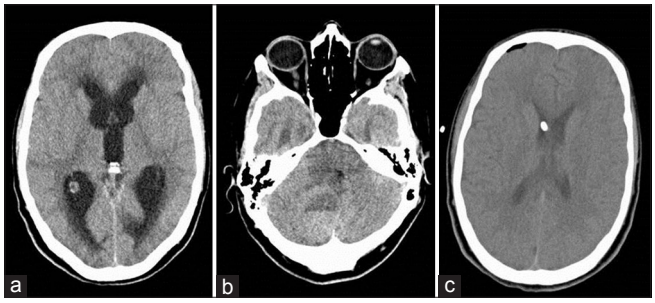

Figure 1: (a and b) Initial computed tomography. (c) Postexternal ventricular drain insertion

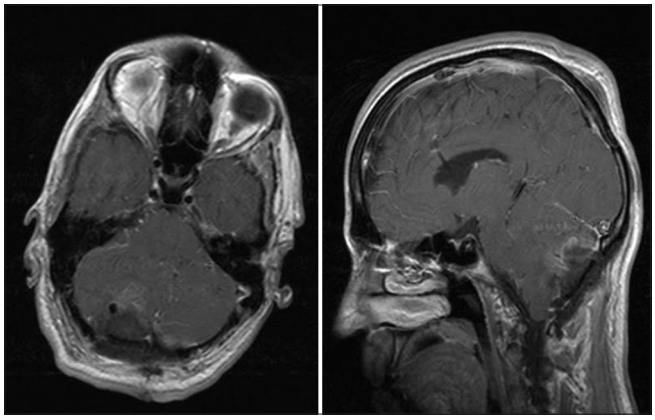

Figure 3: Postoperative magnetic resonance imaging (T1 with contrast)

The turnover (mitosis and necrosis) was high, with Ki67 positivity in 80% of the neoplastic cells [Figure 4c]. Immunohistochemical stains showed weak reactivity of the neoplastic cells for neuron-specific enolase [Figure 4d] and neurofilament proteins and focal reactivity for glial fibrillary acidic protein. There was also positive staining for vimentin. There was no reactivity for synaptophysin of epithelial membrane antigen.

DISCUSSION

Embryonal neoplasms (medulloblastomas, PNETs and atypical teratoid/rhabdoid tumors) are the most frequently encountered primary brain neoplasms in children aged between 0 and 4 years. Medulloblastomas account for 13% of all brain tumors in children aged between 0 and 14 years, 4% in children between 15 and 19 years and 2% in young adults between 20 and 34 years. In adults (defined as age >18 years), medulloblastoma is a very uncommon neoplasm with a slight male predominance (male–female ratio of 1.28).[3]

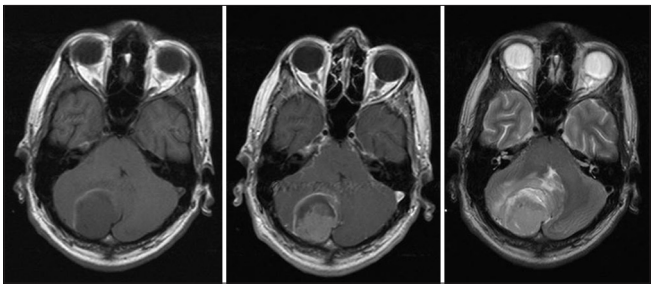

Figure 2: Preoperative magnetic resonance imaging (T1 and T2 images; T1 with contrast)

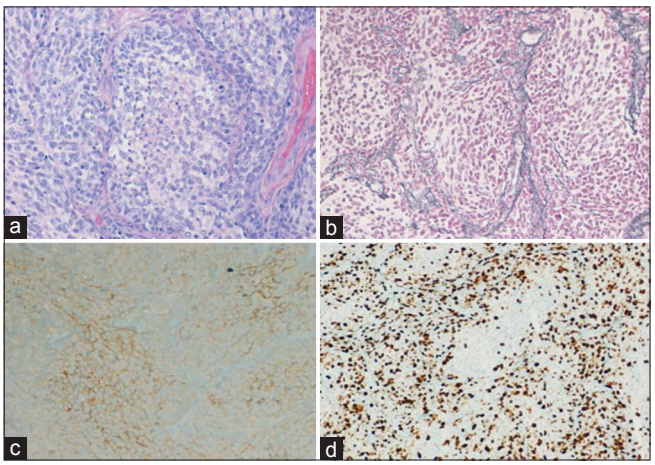

Figure 4: (a) Desmoplastic medulloblastoma, histologic and immunohistochemical features. (a) Note nodules of rounded, primitive neoplastic cells (H and E, ×200). (b) Note reticulin-free nodules encircled by reticulin-rich zones (Reticulin, ×200). (c) Note weak immunoreactivity of neoplastic cells for NSE (IHC, ×200). (d) Note extensive nuclear staining of neoplastic cells for Ki67 (IHC, ×200)

Medulloblastoma is an embryonal neuroepithelial tumor. It is the most frequently encountered primary brain tumor in childhood, representing 25% of all pediatric brain tumors and approximately 30–40% of primary posterior fossa tumors.[4] However, medulloblastoma only accounts for approximately 1% of adult primary brain tumors, the majority of which are diagnosed in the third and fourth decades of life.[5] Medulloblastoma is the third most frequently encountered central nervous system tumor to exhibit systemic metastases after glioblastoma and meningioma.[4]

Most authors agree on a hematogenous route of spread. A shunt tube has also been described as an escape route for tumor cells into the systemic circulation due to the break in the blood–brain barrier, which occurs as a result of surgery to the tumor with the insertion of a cerebrospinal shunt.[4] The tumor commonly arises from the roof of the fourth ventricle, which leads to a local invasion and distant metastases through the CSF.[6]

Medulloblastoma is considered as a Grade IV malignancy by the World Health Organization.[7] Metastasis, young age and residual tumor volume after resection have long been considered as poor prognostic factors in the pediatric population.[8]

There are some peculiar differences between childhood and adulthood medulloblastomas. The desmoplastic form is found more commonly in adulthood (20–40%) than in pediatrics (10%). Furthermore, a location in the hemisphere is more common in adults than in children (approximately 50% vs. 10% of patients, respectively).[9] Moreover, radiological features differ in adults and pediatrics. Homogeneity pre- and post-contrast injection is much less common in adult patients. On CT, it is typically heterogeneous. On MRI, medulloblastoma shows hypointensity and hyperintensity on T1- and T2-weighted images, respectively. After contrast medium injection, the lateral tumors usually show poor enhancement, whereas the vermian tumors show intense enhancement. On magnetic resonance spectroscopy (MRS), medulloblastoma characteristically has an increased taurine peak, detectable at short echo time, an elevated choline peak and a reduced N-acetyl aspartate peak. The increased taurine concentration was found to be an important discriminator of medulloblastomas although it cannot be used as a unique identifier. Some authors have suggested considering the whole MRS pattern for differential diagnosis of posterior fossa tumors in adult patients.[10]

Although similar factors have been studied in adults, the results of the studies are inconclusive. The principle treatments of the lesion in adult patients are surgical resection and complete craniospinal radiotherapy along with a boost of radiation to the bed of the tumor. This treatment regimen yields an approximately 50–60% 5-year survival rate.[4]

According to literature, prognosis is more favorable in medulloblastoma of adults than in children.[11] Few retrospective studies have been published on treatment outcome. Although radiotherapy is recommended, the role of adjuvant chemotherapy is not yet well established.[11] Despite the common use of chemotherapy in pediatric patients, the role of adjunct chemotherapy in adults is still under investigation because of the different neoplastic biology, the rarity of the tumor in adults and a paucity of prospective studies.[4] On the other hand, surgery plays a crucial part and gross total tumor removal is recommended as numerous studies have revealed a close relationship between the extent of resection and prognosis.[12]

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Conflicts of interest

There are no conflicts of interest.

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