EXTRA-AXIAL MEDULLOBLASTOMA IN THE CEREBELLAR HEMISPHERE; A REPORT OF A RARE CASE WITH LITERATURE REVIEW

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Abstract
Extra-axial medulloblastoma is quite rare tumor. This report concern a 13 year old girl who presented with headache, nausea and vomiting of two months duration. Radiological findings demonstrate heterogeneous mass involving the left cerebellar hemisphere and compressing the vermis and the 4th ventricle causing acute hydrocephalus. Patient was treated with ventriculo-peritoneal shunt for hydrocephalus followed by posterior fossa craniectomy and gross total tumor excision. The diagnosis of medulloblastoma is confirmed by histopathology. This is the first reported case of total extra-axially located medulloblastoma in Iraq. Although extremely rare, medulloblastoma should be considered in the differential diagnosis of extra-axial cerebellar hemisphere lesions in children.

Introduction
Medulloblastoma is a common tumor of the posterior fossa in children, representing 20-25% of all pediatric primary malignant central nervous system neoplasms. The tumor often occur in the cerebellar vermis and at the apex of the 4th ventricle. The extra-axial site of this tumor remain a rarity. Previously, 33 cases of medulloblastoma located extra-axially have been reported worldwide but most of these medulloblastomas located at the cerebello-pontine angle (CPA). This is the 1st reported case in Iraq in which a medulloblastoma was entirely located in the cerebellar hemisphere.

Case report
A 13 year old girl presented with nausea and vomiting for 2 months. The initial neurological examination revealed ataxia, dysmetria and advanced bilateral papilledema. Brain CTS showed a well-defined, oval mass in the left posterior fossa, compressing the 4th ventricle causing acute hydrocephalus (Figure 1). An emergency operation was performed, ventriculo-peritoneal shunt, to control the increased intracranial pressure secondary to hydrocephalus. Patient showed improvement after surgery.

Figure 1
Few days later, a brain MRI was performed which revealed presence of an oval, complex, predominantly solid mass with small cystic foci, enhanced heterogeneously following contrast injection, about 4x3.3x2.5 cm in size involving the left cerebellar hemisphere and compressing the vermis and the 4th ventricle with pressure effect on the left side of the pons and the left middle and superior cerebellar peduncles. Ventriculo-peritoneal shunt seen in the right lateral ventricle and well-functioning (Figure 2).

Figure 2

A differential diagnosis of glioma or hemangioblastoma was made, and a continue for the next session of tumor excision 10 days after the 1st operation of CSF shunting. A left lateral sub-occipital infra-tentorial craniectomy was performed for tumor resection. Macroscopically the tumor was well demarcated, semisolid with areas of old hemorrhages, extra-axially located, attached to the adjacent dura of the posterior cranial fossa with invasion of the adjacent cerebellar tissues. Gross total excision was performed. The post-operative course was passed smoothly without complications and patient’s symptoms improved gradually. Histopathological examination reveals diffuse growth pattern of oval cells with hyperchromatic nuclei, large bizarre cells and few rosettes, a picture highly suggesting of medulloblastoma. The slides were revised with other histopathologist who do not know about the first diagnosis, and the result was the same, a classical type of medulloblastoma (Figure 3).

Figure 3

Post-operative contrast-enhanced brain CTS was performed and revealed no evidence of tumor remnant in left posterior fossa (Figure 4). MRI of the spinal cord showed no definite evidence of CSF tumor seedling.
Brain tumors are the second most common malignancy affecting the pediatric population and account for more childhood death than any solid tumors. Medulloblastoma account for 25% of all brain tumors in children and is the most common malignant pediatric tumor of the posterior fossa. Medulloblastoma belong to the family of primitive neuro-ectodermal tumors. The cell of origin is still controversial, it may originate either from germinal cells or their remnant situated at the end of the posterior medullary velum or from the remnant of the external granular layer. Kadın et al suggested that medulloblastoma arise from germinal cells (or their remnant) anywhere along their migratory pathway, and because the migratory process normally proceeds in a lateral direction, a relatively higher frequency of laterally situated tumor should be observed in adults.

Medulloblastoma have long been classified into two chief histological variants, classical and desmoplastic. Recent update to the world health organization classification of CNS tumors have modified the classification of medulloblastoma. The revised classification considered all medulloblastomas to be WHO grade IV tumors. The five subtypes of major histological variants recognized include the classical, desmoplastic/nodular, extensive nodularity medulloblastoma, anaplastic and large cell subtype. The relationship between the histological type of medulloblastoma and their location is still uncertain and should be further studied, but many studies suggested that desmoplastic type have been reported to occur more frequently among extra axial tumors. In this case a classical type was found which is quite unusual regarding the site of the tumor, but it goes with the age of the patient since the classical type is more common in children while desmoplastic type seen more commonly in adult and most of the previously reported extra axial tumors seen in adults.

The clinical presentation of children with medulloblastoma is variable but most patients presented with short history of progressive symptoms which are presented for less than 3 months in 75% of cases. Associated hydrocephalus is responsible for a significant proportion of clinical presentation, as in this case. Controversy still exists regarding the initial management of patient with medulloblastoma and hydrocephalus. Some advocate routine shunting of the hydrocephalus (ventriculo-peritoneal shunt or endoscopic third ventriculostomy) as the first step, claiming that there is a lower mortality and morbidity and a better surgical field after the intracranial pressure is relieved with CSF shunting for a period of several days prior to tumor excision. Still others advocate one session surgery with external drainage of CSF plus direct tumor excision.

The policy at our center is to consider CSF shunting few days prior to craniectomy and tumor excision. Lin et al found that 10-40% of cases demonstrate persistent hydrocephalus after posterior fossa tumor resection alone. Due et al found that the low
cure rate of hydrocephalus by tumor resection alone in patients with medulloblastoma and ependymoma, raises the issue of whether these patients would benefit from preoperative CSF shunting.

The surgical plane is always to try to perform a gross total resection of the tumor because there is evidence in the literatures that this result will enhance the likelihood of long term survival of the patients¹⁹.

Chung et al⁴ revised the prognosis of the extra axially located medulloblastomas in the literatures including 33 cases reported in the cerebellopontine angle, among which 4 cases seen in the pediatric age group described by Kumar et al²⁰ and they concluded that there is currently no clear cut consensus as to whether extra axially located medulloblastomas are more aggressive compared to their midline counterparts⁴.

Conclusion

Medulloblastoma is the most common malignant pediatric tumor in the posterior fossa, and usually occur in the midline. Extra-axial occurrence of medulloblastoma is extremely rare, and clinically it is difficult to be differentiated from other extra axil posterior fossa neoplasms. This is the 1st reported case of extra-axial medulloblastoma in Iraq but still these lesions are likely to be under reported owing to the lack of systematic case reports publication by neurosurgeons in Iraq and wing to publication biases. Although rare, medulloblastoma should be considered in the differential diagnosis of extra-axial posterior fossa lesions.

References