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A Case of Choroid Plexus Carcinoma with Lung Metastasis

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I. Introduction :

In United States incidence rate of primary brain and nervous system tumors in adults is estimated to be 23.8 per 100,000 persons (data from 51 cancer registries, 2013 to 2017) [1]. Approximately one-third of tumors are malignant and the remainder are benign or borderline malignant [1,2].The incidence rate in children (aged 0 to 19 years) is much lower (6.1 per 100,000 children), although in children malignant tumors are more[1].The incidence of central nervous system (CNS) tumors in India ranges from 5 to 10 per 100,000 population with an increasing trend and accounts for 2% of malignancies[3,4].The five most frequent brain tumors in India were astrocytoma (47.3%), medulloblastoma (11.4%), craniopharyngioma (9.7%), ependymal tumors (4.8%), and nerve sheath tumors (4.1%)[5]

Carcinoma of the choroid plexus is an uncommon intracranial neoplasm with a particularly virulent course. Around 80% of choroid plexus carcinoma (CPC) arise in children, in whom they constitute 10–20% of choroid plexus tumors [6]. Here we describe a case of choroid plexus carcinoma in a 23-year-old male with lung metastases

Case Description:

A 23-year-old male was evaluated for headache mainly rightly sided of two weeks duration which later on became holocranial. MRI of the brain showed ill-defined solid cystic lesion with moderate perilesional vasogenic edema in right temporoparietal lobe and thalamus with involvement of atria of ipsilateral lateral ventricles causing gross dilation of temporal horn and adjacent splenium of corpus callosum. Mass effect noted in the lesion causing compression of rest of ipsilateral lateral ventricle and effacement of adjacent cortical sulci. Midline shift to left side measuring 9mm and also mass effect with mild displacement of mid brain. Patient underwent right-into-temporoparietal craniotomy and total excision of the tumor. The post operative histopathology report was choroid plexus carcinoma Grade III of lateral ventricle IHC: CK, S 100, vimentin, pancytokeratin and CK 7 positive. Post-op MRI done showed residual lesion in atrium of right ventricle MRI spine done showed enhancing intradural extramedullary lesion in D1&D2 suggestive of metastasis. Patient underwent craniospinal radiation of 54.8Gy/#/30# along with a boost of 36Gy in 18#.

Patient was referred for adjuvant chemotherapy in view of residual disease, as part of work up CT chest was done which showed bilateral lung metastasis. He was started on chemotherapy with ICE regimen Inj.Ifosfomide 2g/m² for three days, Inj.Carboplatin (AUC)6 for one day and Inj.Etoposide 100mg/m² for three days and has completed 2 cycles of same which was given every three weeks, post two cycles of chemotherapy CT chest done showed partial response and was continued on four more cycles, and he is currently asymptomatic and on follow up.

II. Discussion

Choroid plexus carcinoma (CPC) is a rare cause of a hemispheric cerebral tumor arising from lateral ventricles in children. The possible differential diagnosis for such a hemispheric brain tumor includes choroid plexus papilloma (CPP), ependymoma, atypical teratoid rhabdoid tumor, glioma, astrocytoma, and primitive neuroectodermal tumor (PNET). Radio pathological correlation with tissue immunohistochemistry is of essence in differentiating and establishing a confirmatory diagnosis. CPCs are associated with Li-Fraumeni syndrome, Aicardi syndrome, Simian virus 40 (SV40) on the basis of this viral DNA having been identified in up to 50% of cases.

CPCs are neoplasms of neuroectodermal origin corresponding to WHO grade III tumor. CPC account for 15–20% of choroid plexus tumors, but 80% of these malignant tumors are found in children [6]. Due to their rarity, reports on CPC most often focus on single cases or single-institution experiences with a limited number of patients. Only seven previous series have analyzed 8 or more patients in the last 30 years [7]. The extremely low incidence of CPC in children has been a major obstacle in the development of standardized clinical trials with the therapeutic options being based upon expert opinion and case studies. Various management strategies include surgery, chemotherapy, radiotherapy, and autologous hematopoietic cell rescue.

Clinically, this group of tumors tends to cause hydrocephalus and increased intracranial pressure. Bleggi-Torres et al. reported 15 cases of CPC and pointed out that the main symptoms of this tumor are hydrocephalus (62.5%), intracranial hypertension (25%), and convulsion (12.5%) [7]. Neuroradiological features are nonspecific in CPC. Some features may suggest the diagnosis, such as when the tumor invades the parenchyma or presents with metastatic nodules in the third, fourth, or lateral ventricles. But some choroid plexus papillomas also demonstrate adjacent cerebral edema and invasion, whereas some carcinomas do not. All the tumors in the differential diagnosis, including ependymoma, primitive neuroectodermal tumor, astrocytoma, teratoma, and meningioma, which also can have similar imaging characteristics and modern imaging cannot yet accurately define the pathological diagnosis [7,8].

Surgical resection is considered to be the most effective treatment for CPCs. The extent of surgical resection remains the most important factor in determining long-term survival in patients with CPC, but patients treated only with surgery have had a very poor outcome and disease progresses rapidly and patients often die within 1 year. The early use of radiation therapy may extend survival [7,8]. Unfortunately, radiotherapy is not an option in the majority of cases because of the young age of the patients and the size of the field to be irradiated. Chemotherapy contributes to long-term survival, but it cannot prevent recurrence. Current data strongly support the use of combined chemoradiation in patients older than 3 years and chemotherapy alone if the patients are younger, but the total amount of necessary adjuvant treatment and the order in which the modalities are to be used are still controversial. Unfortunately, the incidence of CPC is too low to set up a randomized study assessing radiotherapy or chemotherapy protocols for patients with CPC [8,9].

Achieving gross total resection (GTR) is the most decisive factor for a patient's long-term survival and prognosis. Various studies have determined that patients who have undergone GTR have significantly better survival rates [10,11]. Furthermore, Mallick et al [12]. observed that progression-free survival was significantly higher for patients with GTR in comparison with subtotal resection (60 months versus 11 months) after eliminating the impact of adjuvant therapy. Despite the merits of GTR, it is difficult to achieve complete resection and it has increased morbidity. The large size, high vascularity, diffuse infiltrative nature, and excessive friability of CPC present a formidable challenge for complete resection. In the pediatric population, blood loss may be life threatening as the entire circulating blood volume may be lost during the resection of these vascular tumors. The surgical approach planned should allow good visual access to vascular supply and maximal exposure of the tumor mass. An effective intraoperative surgical strategy is to identify and ligate the feeding choroidal vessel, thus facilitating the en bloc tumor mass removal. In patients with large tumors where the tumor is resected in parts, gentle coagulation of the fronds of the tumor allows for manipulation without excessive bleeding and may reduce the tumor size. If complete resection is not achieved for any reason, a second-look surgery may achieve an GTR, if this surgery is preceded by administration of chemotherapy, which will help to reduce intraoperative bleeding and tumor size, thus allowing for a complete subsequent resection rather than an incomplete resection [13,14,15].

A global consensus on neoadjuvant chemotherapy and regimens is lacking and is yet to be standardized. The following drugs are used in the treatment: carboplatin, etoposide, cyclophosphamide, high-dose methotrexate, and vinca alkaloids [16]. All meta-analyses have focused primarily on the benefits of using chemotherapy, without a focus on a particular regimen or agent. Using multivariate Cox regression survival analysis, Sun et al [17]. confirmed a better prognosis and a significantly better cumulative overall survival in children with CPC receiving chemotherapy alone. However, the implementation of combined chemoradiotherapy had better overall survival than chemotherapy alone [17]. Besides, among patients with

incomplete resection of CPC, chemotherapy was found to significantly improve the overall survival, but in the subgroup with complete resection, chemotherapy did not make an apparent difference [11]. Additional consideration of chemotherapy is given in children younger than 2 years for whom RT is preferably delayed [12]. Neoadjuvant chemotherapy with ICE regimen has been recommended as it improves the margin of tumor resection with a decrease in the incidence of intraoperative blood loss in children [16-20]. The best chemotherapy regimen is yet to be determined, but a combination utilizing platinum and etoposide as backbone is preferred [12].

Adjuvant chemotherapy appeared to be associated with improved survival [11,17]. This is likely to be employed to defer radiation in children as radiation may hinder in the neurocognitive development. A cautionary tale was, however, sounded by the HITT trials which reported excess death when this strategy was used to delay radiation in anaplastic ependymoma of infants. Without formal trials the best chemotherapy regimen remains unknown. Combinations employing Platinum and Etoposide as a backbone might be preferred based on experience with other pediatric brain tumors. Adjuvant chemotherapy is usually offered to all patients but, as has been previously stated, is of particular importance for those younger than 2 years in whom radiotherapy should be delayed

The common pattern of leptomeningeal progression also raises the role of intrathecal chemotherapy. This may cause less systemic side effects, but at the same time is associated with increased neurocognitive dysfunction, especially when combined with radiation. Methotrexate is the most commonly used intrathecal agent although the evidence for its use in CPC was scarce. The same was true for intrathecal radiopharmaceuticals. Clinical details, proliferation index, and whole genome sequencing may help find high risk groups for treatment intensification in the future. Although adjuvant radiation is very reasonably avoided in younger patients, a risk adapted approach with the early administration of adjuvant radiation in patients with a sub total resection seems logical [12].

Second surgery following neoadjuvant ICE chemotherapy led to a high rate of complete or near complete resection. Chemotherapy appears to facilitate second look surgery, in particular through a reduction of intraoperative blood loss. Despite radiation avoidance, the majority of survivors experienced significant neurocognitive impairment [21].

Pattern of Recurrence and Salvage Treatment [12]

At the median follow-up of 10.8 months, 94 patients had documented progression of disease, but only 11 patients had information about the nature of that progression. Out of these 11 patients, 7 (63.6%) had leptomeningeal spread. One patient developed lung and bone metastasis and another had ascites while another developed a second malignancy, Glioblastoma. Salvage treatment could be retrieved for 9 patients only, of whom 5 underwent re-surgery and 4 received salvage radiation with or without chemotherapy

Although this tumor is still associated with a poor prognosis, there has been a slight but significant increase in survival throughout the past decades. Dohrmann and Collias reported a median survival time in a review of 16 children operated on for CPC [18,19]. In 1992, Packer et al. reported 11 patients with CPC with a 45% event-free survival rate and a median progression-free time of 48 months [9]. Girish et al. reported median survival of 58 months for CPCs who underwent gross total excision with adjuvant therapy and of 36 months who had a subtotal resection with adjuvant therapy [3].

III. Conclusion:

We conclude that, when possible, GTR should be a priority in the management of patients with CPC. However, in the event of subtotal resection, neoadjuvant chemoradiotherapy may be given for a second surgery to achieve a complete GTR. Adjuvant radiotherapy and chemotherapy have been shown to improve survival in a recent meta-analysis. In case of metastasis ICE {ifosfomide, carboplatin, etoposide} chemotherapy which is extrapolated from the chemotherapy used in Neoadjuvant setting seems beneficial

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