Neoadjuvant Chemotherapy in Choroid Plexus Carcinoma: Improving Surgical Safety and Resection Outcomes

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Abstract

Choroid plexus carcinoma (CPC) is a rare and highly vascular malignant brain tumour that occurs mainly in children. Complete surgical resection offers the best chance of long-term survival, but this is often difficult due to excessive intraoperative bleeding. Preoperative (neoadjuvant) chemotherapy has been explored as a strategy to shrink the tumour and reduce its vascularity before surgery. In this review, we have summarized the available literature on preoperative chemotherapy in CPC and its effect on surgical outcomes. Reported cases and small series show that platinum- and etoposide-based regimens can decrease tumour size and intraoperative blood loss, allowing safer and more complete resections. However, due to the rarity of CPC and limited data, larger multicentre studies are required to validate these findings and establish standardized treatment protocols.

Keywords: Choroid plexus carcinoma, preoperative chemotherapy, neoadjuvant therapy, gross-total resection, surgical outcomes

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Introduction

Choroid plexus tumours (CPTs) are rare intraventricular neoplasms of epithelial origin, accounting for approximately 1% of all brain tumours and 2–4% of brain tumours in children.[1] Among CPTs, choroid plexus carcinoma (CPC, WHO grade III) represents the most malignant variant, while choroid plexus papilloma (CPP, WHO grade I) and atypical choroid plexus papilloma (ACPP, WHO grade II) constitute the less aggressive forms.[2]

CPPs are generally benign and are associated with favourable outcomes following complete surgical resection, whereas CPCs are highly aggressive lesions with reported five-year survival rates of only about 40%.[3] Due to their marked vascularity and intraventricular location, CPCs often pose significant surgical challenges, and gross total resection (GTR) at initial presentation remains a key prognostic factor for improved outcomes.[3]

Multimodal therapy combining surgery, chemotherapy, and radiotherapy has improved overall survival in CPC, particularly in cases with residual disease.[4,5] However, while adjuvant treatment approaches are well established, the role of preoperative (neoadjuvant) chemotherapy in improving tumour resection and reducing surgical morbidity remains less clearly defined and warrants systematic evaluation. Hence, this review aims to compile and critically evaluate the available literature on the use of preoperative chemotherapy in CPC and its impact on surgical and oncologic outcomes.

Review of Literature

St. Clair et al., published one of the earliest studies on the management of CPC in children. In their paper, they highlighted the major challenges faced during surgery due to the tumour's extreme vascularity and firm attachment to surrounding brain tissue. The authors observed that achieving gross total resection was often difficult and associated with significant blood loss. They suggested that giving chemotherapy before surgery could be beneficial in reducing the tumour's size and vascularity, which might make the surgical procedure safer and allow for more complete tumour removal. This idea set the basis for later research exploring the role of preoperative chemotherapy in CPC.[6]

In 1997, Razzaq and Cohen described their experience with neoadjuvant chemotherapy in hypervascular malignant brain tumours of childhood, which included patients with CPC. They reported that using chemotherapy before surgery led to noticeable tumour shrinkage and reduced intraoperative bleeding. These findings supported the notion that preoperative chemotherapy can improve the chances of achieving a gross total resection (GTR). The authors also emphasized that this strategy may be especially helpful in infants, who are at higher risk of haemodynamic instability and excessive blood loss during surgery.[7]

Araki et al., provided one of the first detailed case reports showing the clinical benefit of preoperative chemotherapy

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in CPC. They reported a one-year-old infant who initially underwent partial tumour removal, followed by interrupted radiotherapy and two cycles of salvage chemotherapy. Subsequent imaging showed significant tumour shrinkage, which allowed a complete resection in a later surgery. The patient remained disease-free for ten months after the final operation. This case demonstrated that preoperative chemotherapy could make subsequent surgeries easier and potentially improve survival outcomes.[8]

Souweidane et al., contributed important quantitative evidence by using computer-assisted volumetric measurements to evaluate the tumour response to preoperative chemotherapy. Their patient, a fifteen-month-old female with histologically confirmed CPC, received three cycles of multi-agent chemotherapy consisting of etoposide, cyclophosphamide, vincristine, and cisplatin. They observed a 29.5% reduction in tumour volume before surgery, which enabled staged complete removal of the tumour. The patient remained disease-free for 31 months. The authors concluded that volumetric measurements could serve as an objective method to evaluate chemotherapy response and optimize treatment regimens for CPC.[9]

Gopal et al., focussed on the challenges associated with CPC surgery and emphasized the morbidity related to extensive resection. They discussed that because these tumours are highly vascular and often disseminated at diagnosis, achieving complete resection can be associated with substantial risks. They mentioned that the use of neoadjuvant chemotherapy to reduce tumour vascularity had been reported, although it was not yet a standard approach at that time. This observation highlighted the need for further studies to define the potential role of preoperative chemotherapy in improving surgical outcomes.[10]

A significant contribution to this field was made by Schneider et al., who reviewed 22 paediatric cases of CPC treated between 1982 and 2013, of which 12 patients received neoadjuvant ICE (ifosfamide, carboplatin, etoposide) chemotherapy. The study found that in all the cases where preoperative chemotherapy was used, either gross total or near-total resection (more than 95% of tumour volume) was achieved. Moreover, the mean blood loss in these patients was only 22% of the total estimated blood volume compared to 96% in those who did not receive chemotherapy. The authors concluded that preoperative chemotherapy significantly reduced intraoperative blood loss and improved both the extent of resection and overall survival.[11]

More recently, a systematic review and institutional series from Texas Children's Hospital (2025) analysed data from 21 studies including 124 patients, 26 of whom had CPC. The study found that 82.3% of patients showed a measurable reduction in tumour size following preoperative chemotherapy, and 78.2% achieved gross total resection. Importantly, no chemotherapy-related surgical complications were reported. The authors concluded that preoperative chemotherapy is a safe and effective approach to improve the safety and completeness of tumour resection in paediatric brain tumours, including CPC.[12]

In addition to clinical findings, Liu et al., contributed valuable molecular insights through the SJYC07 trial. Although chemotherapy in this study was administered postoperatively, several patients underwent second-look surgeries after induction chemotherapy, demonstrating the concept of tumour cytoreduction before definitive resection. The study also identified frequent TP53 gene alterations—many of which were germline mutations associated with Li-Fraumeni syndrome—that were linked with poorer outcomes. These molecular findings suggest that genetic factors may influence tumour behaviour and response to chemotherapy, providing a new direction for future research into individualized treatment approaches for CPC.[13]

In 2025, the European Society for Paediatric Oncology (SIOPE) Brain Tumour Group published updated guidelines for the management of CPC (Brain & Spine, 2025). They emphasize maximal safe resection as the main treatment, supported by multi-agent chemotherapy, most often including platinum- and etoposide-based regimens.[14] The guidelines also recognize that preoperative (neoadjuvant) chemotherapy can be used when gross-total resection is not possible at first because of tumour vascularity or patient condition. They further highlight the importance of molecular testing, especially TP53 mutation status, to guide prognosis and tailor therapy. These recommendations align with previously published reports, showing that neoadjuvant chemotherapy can make surgery safer and more effective in difficult CPC cases.[14]

Conclusion

Preoperative chemotherapy appears to be a useful approach to improve the safety and completeness of CPC surgery. Studies consistently show decreased tumour vascularity, less blood loss, and higher chances of total resection after neoadjuvant therapy. However, since CPC is very rare and most published data are from small, retrospective studies, it is difficult to form a universal protocol. Future multicentre and prospective studies are

needed to identify the best chemotherapy combinations, timing, and patient selection, so that treatment for this aggressive paediatric tumour can become safer and more standardized.

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