Diffuse Midline H3 K27-Altered Gliomas in the Spinal Cord: A Systematic Review

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Background: Gliomas account for 80-90% of all intramedullary spinal cord tumors (IMSCTs). Though rare compared to brain tumors, spinal cord gliomas can cause significant morbidity and mortality. Diffuse midline gliomas (DMGs) with H3 K27M-mutation, first introduced in the 2016 WHO classification, are high-grade tumors with aggressive behavior and poor prognosis. The 2021 updated WHO classification renamed them "diffuse midline glioma, H3 K27-altered" to include other molecular changes. Limited single-institution data on spinal cord DMGs (DMG-SCs) hinder comprehensive understanding and optimal treatment protocols. In this review, we summarize clinical and molecular features, management strategies, and survival impact in patients with DMG-SCs.

Project Methods: A systematic review was performed following the (PRISMA) guidelines. PubMed, Ovid EMBASE, Scopus, and Web of Science were searched. Clinical characteristics, treatment protocols, and outcomes were analyzed.

Results: A total of 26 studies with 259 patients were included. Most patients were male (63%), diagnosed at a mean age of 32 years (range, 4-72), and tumors were predominately located in the cervical (32%) or thoracic (43%) regions of the spinal cord. Primary management included surgical resection (97%), radiotherapy (78%), and chemotherapy (62%). Most common combination of treatment included surgical resection, radiotherapy, and chemotherapy (47%). The mean overall and progression free survival were 25 (range, 0.1-48) and 14 (range, 0.1-25) months, respectively. Gene alterations included p53 mutation (61%), loss of ATRX (46%), Olig-2 positive (100%), and GFAP positive (80%). The mean Ki-67/MIB-1 was 23% (12-40%).

Conclusion/Impact: DMG-SCs affect mostly the adult population and appear to resemble adult DMGs in terms of molecular features, management, and prognosis.
Extracranial Meningioma Metastasis: A Systematic Review of Clinical Characteristics, Management Strategies, and Outcomes

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Background: Meningioma is the most common type of intracranial neoplasm, accounting for approximately 40% of all primary brain tumors. Although these tumors are usually benign and slow-growing, extracranial metastasis can occur in less than 1% of cases. Due to the rarity, diagnosis can pose a challenge. In this systematic review, we summarize and analyze patient demographics, clinical characteristics, management strategies, and outcomes of patients with extracranial meningioma metastasis.

Project Methods: A systematic review was performed following the (PRISMA) guidelines. PubMed, Ovid EMBASE, Cochrane, Scopus, and Web of Science databases were searched. Clinical characteristics, management, and outcomes were analyzed.

Results: A total of 127 studies with 164 patients were included. There were 51% males and mean age of primary tumor diagnosis was 48 years (range, 8-91). Primary tumors were mostly located on the convexity of the brain (52%) and WHO grade 1 (38%) or grade 2 (37%). Histological findings were predominantly atypical (37%). Mean number of intracranial recurrences was 2 (range, 0-7) and occurred in 81% of cases. Average time between primary tumor and the first extracranial metastasis was 103 months (range, 2-450). The top three most common locations of metastases were the lungs (39%), spine (15%), and liver (12%). Most often, there was no change in grade (68%) from the primary tumor to the first metastasis. Gross total resection of the primary tumor was achieved in 76% of cases. Mean survival from primary diagnosis and survival from first metastasis was 118 and 31 months, respectively.

Conclusion/Impact: Mechanisms by which extracranial meningioma metastasis occur are still unclear, though do not appear to involve evolution into a more aggressive histologic type in most cases. In a patient with a history of intracranial meningioma recurrence and symptoms of lung, spine, or liver, dysfunction, extracranial meningioma metastasis should be considered within the differential.
Intraventricular Ependymoma in Pediatric Patients: A Systematic Review of Demographics, Clinical Characteristics, and Outcomes

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Background: Intraventricular neoplasms are rare occurrences observed in 5 – 7% of all primary pediatric brain tumors. Pediatric intraventricular ependymomas are a complex subset of these tumors, poorly discussed across the current literature. Although surgery is generally the accepted treatment of choice, information on clinical course and outcomes is limited to heterogeneous case reports and small case series focusing on specific histologic subtypes or ventricular locations. We conducted a systematic review on pediatric intraventricular ependymomas to survey the patient population, tumor characteristics, management strategies, and associated outcomes.

Project Methods: PubMed, Scopus, Web-of-Science, and Cochrane were searched upon the PRISMA guidelines to include studies reporting pediatric patients with intraventricular ependymomas. Clinical characteristics, treatment protocols, and outcomes were analyzed.

Results: A total of 9 studies with 70 patients were included. Most patients were male (54%), diagnosed at a mean age of 7 years (range, 0.2-17), and frequently exhibited nausea and vomiting (38%), headache (31%), and ataxia (25%). Tumors were predominantly located in the fourth ventricle (79%) and most tumors were WHO grade 2 (73%). Mean tumor volume was 3 cm³ (range, 0.1-13.2). Management included surgical resection (96%), radiotherapy (87%), and chemotherapy (38%). Gross total resection was achieved in 69% of cases. Cranial nerve deficit was the most common post-surgical complication (71%). Most common combination of treatment included surgical resection and radiotherapy (53%). Mean overall survival was 50 months in these patients.

Conclusion/Impact: Pediatric intraventricular ependymomas are rare tumors with limited information on management strategies. The mainstay of treatment is complete surgical resection. Compared to ependymomas, intraventricular ependymomas appear to have a worse overall prognosis.