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Gliomas

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Continuing Education Activity

Gliomas are a diverse group of tumors originating from glial cells in the brain and spinal cord, representing the most common primary brain tumor within the central nervous system. In the United States, there are 6 cases of gliomas diagnosed per 100,000 individuals every year. These tumors vary widely in their aggressiveness and prognosis, ranging from low-grade pilocytic astrocytomas to highly malignant glioblastomas. Advances in molecular and genetic research have led to improved diagnostic accuracy and the development of targeted therapies. Despite these advancements, the diffuse and infiltrative nature of high-grade gliomas poses ongoing challenges in treatment and management.

This activity reviews the pathophysiology, clinical presentations, and diagnostic techniques of glioma and provides the knowledge and skills to diagnose, treat, and manage patients with gliomas. Participants gain insights into current and emerging treatment options, including surgery, radiation therapy, and chemotherapy, as well as the latest research findings and multidisciplinary approaches to improving patient outcomes. The activity also highlights the role of the interprofessional healthcare team in supporting comprehensive care for affected patients.

Objectives:

- Identify early signs and symptoms of gliomas, such as new-onset headaches, seizures, cognitive deficits, or focal neurological deficits, to facilitate timely diagnosis and intervention.
- Differentiate between various types of gliomas based on histopathological characteristics, molecular markers, and imaging findings to guide appropriate treatment strategies.
- Implement evidence-based treatment protocols tailored to the specific type and grade of glioma, incorporating the latest advances in medical research.
- Evaluate strategies to enhance care coordination and communication among interprofessional team members to improve outcomes for patients affected by gliomas.

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Introduction

Gliomas are the most common primary brain tumors of the central nervous system (CNS). In the United States, there are 6 cases of gliomas diagnosed per 100,000 individuals every year.[1] Gliomas can be well-circumscribed or diffusely infiltrative tumors that affect all areas of the brain. Gliomas range from grade 1 tumors (ie, pilocytic astrocytomas) to grade 4 tumors (ie, glioblastoma), per the World Health Organization grading system.

In the past, diffuse gliomas were classified and subtyped based on histopathology, such as diffuse astrocytoma, oligodendrogliomas, or mixed gliomas/oligoastrocytomas. More recently, gliomas have been classified based on molecular and genetic markers.[2] These advances have led to more specific prognostic and therapeutic targets.

Etiology

There are 3 common types of gliomas, which are classified based on their phenotypic cell characteristics: astrocytomas, ependymomas, and oligodendrogliomas. These gliomas are further classified into low-grade, atypical/anaplastic, or high-grade tumors based on cell morphology, mitotic activity, and molecular markers. The WHO grading system utilizes molecular markers and has been shown to have significant prognostic and therapeutic implications.[3]

- **Astrocytomas**: Originate from astrocytes and can be encapsulated, preserving clear borders between normal brain tissue and tumor cells or infiltrative—indicating a higher-grade lesion. Low-grade tumors are more common in children, while high-grade neoplasms are more common in adults.
- **Oligodendrogliomas**: These gliomas originate from oligodendrocytes and are less infiltrating than astrocytomas. They are commonly found in middle-aged adults.
- **Ependymomas:** These gliomas originate from ependymal cells that line the ventricular cavities and the central canal of the spinal cord; they are more common in young children.

Epidemiology

There are an estimated 80,000 to 90,000 newly diagnosed cases of primary brain tumors each year in the United States, with approximately 25% being gliomas. The total number of glioblastoma cases diagnosed each year is about 12,000 (approximately 15% of the total of newly diagnosed brain tumors and roughly 50% of all malignant brain tumors).[4] Gliomas most often present in patients without any relevant history; however, some patients may have tumor-predisposition syndromes. The phacomatoses such as neurofibromatosis type 1 and neurofibromatosis type 2 predispose patients to astrocytomas and ependymomas, respectively. Furthermore, patients with tuberous sclerosis are predisposed to having subependymal giant cell astrocytomas. Li-Fraumeni is another tumor predisposition syndrome characterized by mutations in p53 and an increased risk of IDH-wild type high-grade astrocytic gliomas.[5]

Pathophysiology

Gliomas are subdivided into astrocytoma, oligodendroglioma, or glioblastoma based on 2 molecular markers: isocitrate-dehydrogenase (IDH) and 1p/19q co-deletion. IDH mutant tumors have elevated levels of 2-hydroxyglutarate (2-HG) and decreased production of nicotinamide adenine dinucleotide phosphate that can lead to increased histone methylation and deoxyribonucleic acid hypermethylation. The increase in 2-HG can also lead to local immune responses and glioma-induced epilepsy.[6]

According to the WHO 2021 recommendations for central nervous system tumors, astrocytomas are now diagnosed solely by IDH mutations.[7] Oligodendrogliomas, by definition, have IDH mutations and 1p/19q co-deletions. New to the 2021 recommendations, glioblastomas are now only IDH-wildtype tumors and no longer classified as IDH-mutant. Several other biomarkers have been identified in gliomas, such as alpha-thalassemia X-linked intellectual disability mutations and mutations in *TP53*. Another molecular signature, CDKN2A/CDKN2B homozygous deletions, has been shown to have a worse prognosis in IDH-mutation astrocytomas.[8] Results from some epidemiological studies suggest that ionizing radiation can increase the chance of developing gliomas.[9]

Histopathology

Gliomas form a heterogeneous group of central nervous system tumors. For decades, histologic diagnosis of these tumors provided a valuable foundation for assessing prognosis and guiding

therapeutic management. However, it is now evident that understanding the molecular underpinnings of glial neoplasms offers a more precise and robust classification.[10]

History and Physical

The most common symptoms of gliomas are headaches, seizures, nausea, vomiting, and focal neurological deficits. The neurological examination of these patients can be normal or present with varying degrees of focal weakness, sensory deficits, or, in a more severe situation, altered mental status due to a mass effect resulting from peritumoral edema. Patients can also develop acute, obstructive hydrocephalus as in cases of large posterior fossa pilocytic astrocytomas. Hydrocephalus can also be seen with diffuse-midline gliomas arising in the pons, tectal plate gliomas, and subependymal giant cell astrocytomas. Patients with optic-pathway gliomas can have varying degrees of visual loss depending on the extent of the disease. Although most patients are symptomatic at diagnosis, approximately 3% to 10% of low-grade gliomas are found incidentally upon work-up for other conditions.[11][12]

Evaluation

Imaging diagnostics are appropriate for evaluating suspected brain tumors and include the following:

- **Computed tomography (head)**: When patients present to the emergency department for evaluation, their initial imaging is usually a computed tomogram (CT). A head CT is an excellent initial study to evaluate for acute intracranial findings such as hemorrhage, edema, and asymmetry in the brain tissue.
- **Computed tomography (chest, abdomen, and pelvis)**: Patients with newly diagnosed lesions suggestive of a brain tumor should undergo a metastatic workup with advanced imaging in search of a primary source (ie, lung, breast, colon, renal).
- **Magnetic resonance imaging (brain)**: This is the most sensitive study to determine the characteristics of brain tumors. Low-grade tumors usually do not enhance, while high-grade tumors have varying degrees of enhancement. In addition, magnetic resonance imaging (MRI) can better assess the degree of cerebral edema. An MRI will facilitate the treatment plan and response to surgery, radiation, and chemotherapy.
- **Magnetic resonance imaging (spine):** Patients who have cranial lesions suggestive of an ependymoma should undergo contrasted imaging of the entire spinal axis to evaluate for drop metastases.
- **Diffusion tensor imaging (brain):** Patients undergoing surgical workups may benefit from tractography for surgical planning.
- **Functional magnetic resonance imaging:** If lesions arise near the eloquent cortex, a functional MRI of the brain may aid in determining the precise location of language or motor function.

Treatment / Management

The WHO classification of gliomas is used to guide treatment. As indicated in the classification, most patients will require surgical intervention via maximally safe resection or stereotactic-guided biopsy.[13]

2021 WHO Classification of Central Nervous System Gliomas

- **Adult-type diffuse gliomas**
  - Astrocytoma, IDH-mutant (WHO grades 2-4)
  - Oligodendroglioma, IDH-mutant, 1p/19 co-deleted (WHO grades 2-3)
  - Glioblastoma, IDH wild-type (WHO grade 4)
- **Pediatric-type diffuse low-grade gliomas**
  - Diffuse astrocytoma, MYB or MYBL1 mutated (WHO grade 1)
  - Angiocentric glioma (WHO grade 1)
  - Polymorphous low-grade neuroepithelial tumor of the young (PLNTY)
  - Diffuse low-grade glioma, MAPK pathway altered
- **Pediatric-type diffuse high-grade gliomas**
  - Diffuse midline glioma, H3K27 altered (WHO grade 4)
  - Diffuse hemispheric glioma, H3G34 mutant (WHO grade 4)
  - Diffuse pediatric-type high-grade glioma, H3 wild-type, and IDH wild-type (WHO grade 4)
  - Infant-type hemispheric glioma
- **Circumscribed astrocytic gliomas**
  - Pilocytic astrocytoma (WHO grade 1)
  - High-grade astrocytoma with piloid features
  - Pleomorphic xanthoastrocytoma (WHO grades 2-3)
  - Subependymal giant cell astrocytoma (WHO grade 1)
  - Chordoid glioma (WHO grade 2)
  - Astroblastoma, MN1 altered
- **Ependymal tumors**
  - Subependymoma
  - Myxopapillary ependymoma
  - Spinal ependymoma, MYCN amplified
  - Spinal ependymoma
  - Posterior fossa ependymoma, group PFA
  - Posterior fossa ependymoma, group PFB
  - Posterior fossa ependymoma
  - Supratentorial ependymoma, ZFTA fusion-positive
  - Supratentorial ependymoma, YAP1 fusion-positive
  - Supratentorial ependymoma [7]

Treatment of Gliomas

Surgery: Surgical resection is the mainstay of treatment with a goal of maximal safe resection, depending on tumor grading and location.

- **Grade I:** Most of these gliomas are surgically curable; however, in some cases, such as optic-pathway gliomas, surgical resection may be limited.
- **Grade II:** Current acceptable practices include a maximal safe resection and the possibility of gross total resection, with radiographic follow-up.
- **Grade III:** Maximal safe resection, concomitant chemoradiation, and radiographic follow-up for recurrence is an acceptable treatment.
- **Grade IV:** Maximal safe resection, concomitant chemoradiation, and radiographic follow-up for recurrence is an acceptable treatment. In select cases, as in diffuse midline gliomas, surgery is often limited to a stereotactic biopsy for molecular characterization.

Chemoradiation: Currently, the Stupp protocol is the standard of care for high-grade gliomas.[14] The protocol consists of radiotherapy and concomitant chemotherapy with 2Gy per day fractions for a total of 60Gy over 6 weeks and temozolomide.

Treatments for Recurrence: Options for recurrent gliomas include re-operation, targeted therapy such as angiogenesis inhibitors [15], or tumor-treating fields.[16]

Other Treatments: Patients with gliomas are prone to seizures, malignant edema, and complication-related immobility. Therefore, these patients may need antiepileptic medications, deep venous thrombosis prophylaxis, and steroids before, during, and after the course of treatments to treat cerebral edema. In some cases, such as diffuse midline gliomas arising in the pons and ependymomas of the fourth ventricle, patients may require treatment of hydrocephalus via ventriculoperitoneal shunting [17] or endoscopic third ventriculostomy.[18][19]

Differential Diagnosis

When evaluating a patient with a suspected glioma, it is crucial to consider a range of differential diagnoses to ensure accurate diagnosis and appropriate treatment. Gliomas can present with symptoms that overlap with various other neurological conditions, including other primary brain tumors, metastatic lesions, and non-neoplastic disorders. Identifying these potential differentials helps formulate a comprehensive diagnostic approach and tailor patient management plans effectively.

The differential diagnoses for glioma include the following:

- Abscess, bacterial or fungal
- Cerebritis/encephalitis
- Tumefactive demyelination
- Gliosis
- Infarct
- Metastasis
- Focal cortical dysplasia

Surgical Oncology

Several techniques have been used to increase the extent of resection of gliomas, including intraoperative MRI and fluorescein-guided resections.[20][21] Ongoing clinical trials are investigating novel surgical adjuncts, such as laser interstitial thermal therapy, to further improve the efficacy and safety of glioma surgery.

Radiation Oncology

Postoperative radiation therapy aims to eliminate residual tumor cells, reduce recurrence risk, and prolong survival. Techniques such as intensity-modulated radiation therapy and stereotactic radiosurgery allow for precise targeting of the tumor while sparing healthy brain tissue, minimizing adverse effects.

Landmark studies, such as those conducted by the Radiation Therapy Oncology Group, have established the benefit of radiation in extending survival for glioma patients. Despite its efficacy, radiation therapy can lead to adverse effects, including cognitive decline, radionecrosis, and secondary malignancies. Ongoing research and clinical trials are focused on refining radiation delivery methods and combining radiation with novel therapies, such as immunotherapy and targeted agents, to enhance treatment outcomes and reduce adverse events.

Medical Oncology

As many pediatric low-grade gliomas have genomic alterations in the mitogen-activated protein kinase pathway, studies have focused on targeted therapies involving this pathway. The phase 2 FIREFLY-1 trial assessed the efficacy of the RAF inhibitor tovorafenib in patients who suffered from relapse or refractory low-grade gliomas with BRAF alterations.[22] This study demonstrated an overall response rate of 67% and 51% when assessed using the Response Assessment in Neuro-Oncology (RANO) for high-grade gliomas (HGG) and Response Assessment in Pediatric Neuro-Oncology (RAPNO) criteria, respectively. Furthermore, the median time to treatment response was 3.0 months and 5.3 months per RANO-HGG and RAPNO criteria, respectively. These findings provide encouraging evidence for another treatment option for patients with relapsed or refractory low-grade gliomas. Several other studies have shown promising results with chemotherapy-directed treatment for diffuse midline gliomas, H3K27M altered.[23][24]

Prognosis

The prognosis of patients with gliomas depends on several factors, as follows:

- Age of the patient
- Comorbidities
- Grade and location of the tumor
- Presence of hydrocephalus
- Response to adjuvant therapy
- Extent of surgical resection

Low-grade gliomas (WHO grade I and II) generally have a more favorable prognosis with longer survival rates, particularly when complete surgical resection is achievable. High-grade gliomas (WHO grade III and IV), including glioblastomas, have a much poorer prognosis due to their aggressive nature and high propensity for recurrence. Advances in molecular profiling have led to more precise prognostic information, identifying markers such as IDH mutations and 1p/19q co-deletions that are associated with better outcomes. Despite these advancements, the overall

survival for high-grade gliomas remains limited, highlighting the urgent need for continued research and development of new therapeutic strategies.

Complications

Gliomas, particularly high-grade forms like glioblastomas, pose numerous complications that significantly impact patient outcomes and quality of life. These complications arise from the tumor's infiltrative nature, which can lead to extensive neurological deficits. Additionally, treatment-related adverse effects further complicate management and recovery.

Complications of gliomas include the following:

- Increased intracranial pressure
- Seizures
- Hydrocephalus
- Brain herniation
- Hemorrhage into the tumor
- Local spread
- Spinal metastases
- Death

Postoperative and Rehabilitation Care

Postoperative and rehabilitation care for patients with gliomas is critical to optimizing recovery and improving quality of life. After surgery, patients require close monitoring for complications such as infection, bleeding, and neurological deficits. Early initiation of rehabilitation is essential, involving a multidisciplinary team that includes physical therapists, occupational therapists, speech therapists, and neuropsychologists.

Rehabilitation aims to address the specific functional deficits resulting from the tumor and its treatment, such as motor weakness, speech difficulties, and cognitive impairments. Personalized rehabilitation programs focus on regaining independence, enhancing physical capabilities, and supporting mental health. Additionally, ongoing follow-up care is crucial to monitor for tumor recurrence, manage adverse effects of long-term treatment, and provide supportive care.

Consultations

Patients with gliomas benefit from consultations with a multidisciplinary team of specialists to ensure comprehensive and personalized care. Neuro-oncologists play a central role in coordinating treatment plans, incorporating input from neurosurgeons who assess surgical options and perform tumor resections when feasible. Radiation oncologists provide expertise in radiation therapy planning and delivery, aiming to optimize treatment outcomes while minimizing side effects. Neuro-radiologists contribute through advanced imaging techniques for accurate diagnosis and monitoring of tumor response. Neuropathologists analyze tissue samples to confirm diagnoses and provide molecular insights that guide treatment decisions. Additionally, supportive care from psychologists, social workers, and palliative care specialists helps address emotional, social, and quality-of-life concerns throughout the treatment journey. This collaborative approach ensures that glioma patients receive integrated care that addresses the complexities of their condition.

Deterrence and Patient Education

Deterrence and patient education are crucial components in managing gliomas, aiming to improve patient outcomes and quality of life. While the exact causes of gliomas are not well understood, certain lifestyle modifications and preventive measures may help reduce risk factors associated with brain tumors. Educating patients about the importance of regular medical check-ups, avoiding exposure to environmental carcinogens, and maintaining a healthy lifestyle can be beneficial.

Additionally, patients and their families should be informed about the symptoms of gliomas, such as persistent headaches, seizures, and neurological deficits, to ensure early detection and prompt medical attention. Providing comprehensive education on treatment options, potential adverse effects, and the importance of adherence to therapy can empower patients to actively participate in their care. Regular follow-up visits, taking medications on time, and driving restrictions must be addressed. Support resources, including counseling and support groups, can also play a vital role in helping patients cope with the diagnosis and treatment of gliomas.

Pearls and Other Issues

Understanding the nuances of managing gliomas requires integrating clinical insights and evidence-based practices. Clinical pearls that guide healthcare professionals in navigating the complexities of diagnosis, treatment, and care for patients with gliomas include the following:

- **Early diagnosis and imaging**: Prompt recognition of symptoms like persistent headaches, seizures, and neurological deficits is crucial for early diagnosis.
- **Histopathology versus molecular profiling**: Incorporate histopathological evaluation and molecular profiling for accurate diagnosis and prognostication.
- **Maximal safe resection**: Strive for maximal safe surgical resection to improve outcomes, balancing tumor removal with preservation of neurological function.
- **Combination therapy**: For optimal management, utilize multimodal treatment approaches combining surgery, radiation therapy, and chemotherapy (eg, temozolomide).
- **Monitoring and surveillance**: Regular imaging and clinical assessments are essential to monitor disease progression and response to treatment.
- **Supportive care**: Address symptom management and quality of life issues through supportive care interventions, including rehabilitation and palliative care.
- **Clinical trials**: Consider enrollment in clinical trials to access novel therapies and contribute to advancing treatment options.
- **Patient and family education**: Provide comprehensive education on the disease process, treatment options, and available support resources to empower informed decision-making.
- **Long-term follow-up**: Establish a structured follow-up plan to monitor for recurrence, manage treatment-related complications, and support survivorship care.
- **Multidisciplinary team approach**: Collaborate closely with neuro-oncologists, neurosurgeons, radiation oncologists, nurses, social workers, and other specialists to deliver comprehensive and coordinated care.

Enhancing Healthcare Team Outcomes

Enhancing patient-centered care, outcomes, patient safety, and team performance related to gliomas requires a coordinated effort across various healthcare professionals, each contributing unique skills and perspectives. Physicians, including neuro-oncologists and neurosurgeons, apply

specialized knowledge in diagnosing, staging, and managing gliomas. Advanced practitioners, such as nurse practitioners and physician assistants, play crucial roles in patient assessment, treatment planning, and symptom management, ensuring continuity of care. Nurses provide frontline support, administer treatments, monitor patients, and provide education about their condition and care plans. Pharmacists contribute by managing complex medication regimens, ensuring drug efficacy and safety, and monitoring for adverse effects. Social workers help patients navigate the healthcare system, connect with community resources, and access financial assistance or insurance coverage.[25]

Responsibilities are shared among healthcare team members, emphasizing clear communication, collaboration, and respect for each profession's scope of practice. Interprofessional communication fosters cohesive teamwork, enabling seamless care transitions and shared decision-making. Care coordination ensures that patients receive timely interventions, access to supportive services, and holistic management addressing physical, emotional, and psychosocial needs.

Review Questions

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