

Pilocytic Astrocytoma

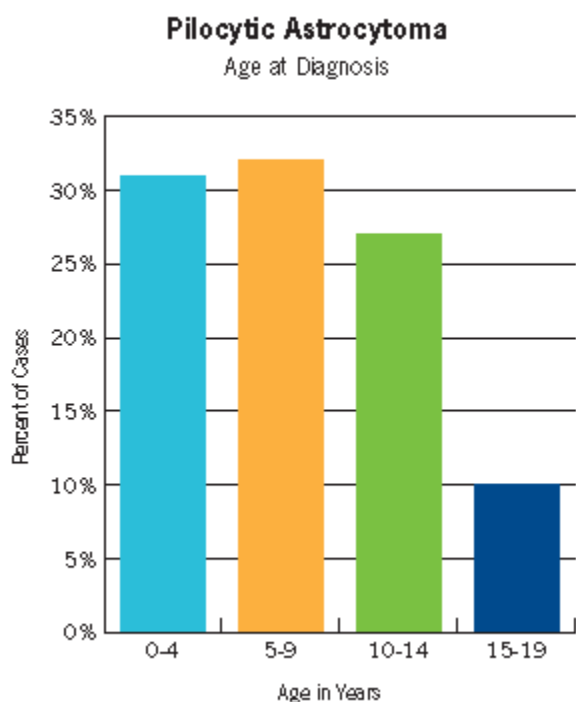
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Tumors of the brain and spinal cord are among the most common malignancies in pediatrics. Annually, these account for approximately 20-25 percent of all new cancer diagnoses in children. Advances in treatment and supportive care have led to improved outcomes, but overall central nervous system malignancies represent a disproportionate amount of morbidity and mortality, and are the leading cause of cancer-related deaths in the pediatric population. Pilocytic astrocytomas (PA) are benign tumors that can occur throughout the brain. Typically well-circumscribed and slow growing, these tumors represent 20-30 percent of all CNS malignancies. Typically, they are graded as WHO I. From 2004-2007, there were a total of 2,500 new cases of pilocytic astrocytomas in the United States. There is no significant difference in incidence of disease by race, age or sex.

Neurofibromatosis type 1 (NF-1) is associated with an increased risk for developing PA. The treatment of these tumors in patients with NF-1 varies significantly because patients with NF-1 are at a greater risk of radiation-associated secondary malignancies. Other genetic risk factors for PA have not been identified. Cytogenetic abnormalities in these tumors are uncommon and the etiology remains unknown.

Clinical presentation

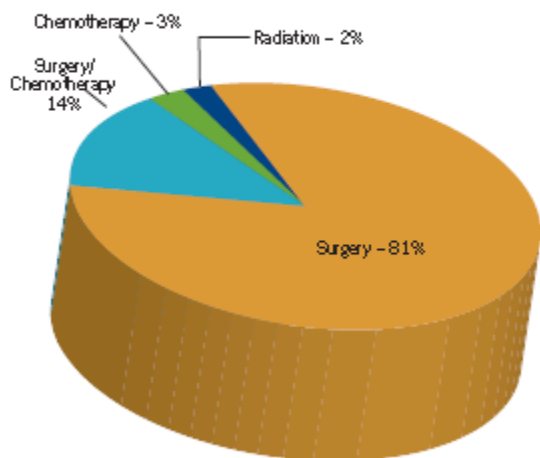
Pilocytic astrocytomas present in a varied fashion usually determined by location. Tumors in the midbrain or cerebellum typically present with symptoms of obstructive hydrocephalus, including irritability, vomiting and progressive headache. Tumors in the supratentorium can result in focal signs, including unilateral extremity weakness, gait abnormality or visual disturbances. Rarely, a form of PA that develops in the hypothalamus in infants can cause diencephalic syndrome, which is characterized by weight loss, irritability and nystagmus, but normal linear growth.



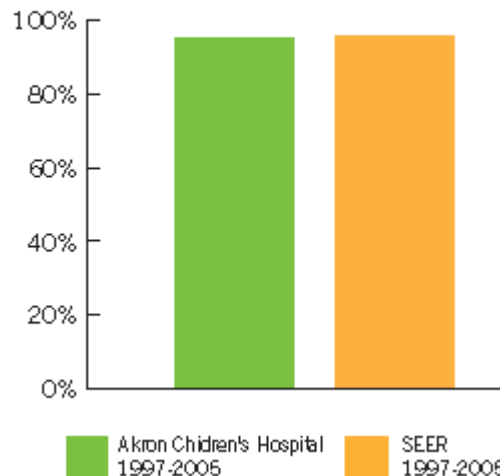
Treatment

The mainstay of treatment for PA is complete surgical excision, which is usually curative. Patients whose tumors cannot be removed completely may need additional treatment including radiation therapy or chemotherapy. The current treatment for partially resected or completely unresected tumors is controversial, and the optimal regimen is dictated by age. Chemotherapeutic agents including vincristine, carboplatin or carmustine have been shown to prolong survival in children younger than age 10 in whom radiation may cause significant morbidity. Adolescents and children older than 10 are better able to tolerate radiation, which is also effective in prolonging survival. In general, patients with PA have excellent survival, and treatment regimens are tailored to each patient, taking into account the morbidity that accompanies each type of therapy.

Pilocytic Astrocytoma Treatment Breakdown



Overall Five-Year Relative Survival Pilocytic Astrocytoma



The Akron Children's Experience

In the period from 1995-2010, 59 new cases of pilocytic astrocytoma were treated at Akron Children's Hospital. Age at diagnosis ranged from 6 months to 25 years old. The ratio of male to female patients was approximately 1:1, and more than 90 percent of patients were Caucasian. The remaining were African-American. Two of the patients presented with a spinal lesion in addition to the intracranial lesion, while the rest presented with tumors isolated to the brain. The majority (81 percent) of all patients were treated with surgical resection alone. Fourteen percent received surgery and chemotherapy, and less than 5 percent received either radiation or chemotherapy alone. Most (95 percent) of the children treated at Akron Children's Hospital from 1997-2005 are currently alive. This is statistically consistent with information available in the national SEER database that shows a five-year overall survival rate of 96 percent during the same time period. This excellent survival rate, consistent with the best pediatric centers nationwide, reflects the high level of care provided to patients at Akron Children's Hospital.

References

1. Central Brain Tumor Registry of the United States (CBTRUS) Report, February 2011.
2. SEER cancer statistics review, 1995-2007.
3. Stuart H. Orkin, Oncology of infancy and childhood 2009.