COMMENT. Epidermal nevus syndrome (or Solomon syndrome) is characterized by various epidermal nevi, including ichthyosis, acanthotic, and sebaceous, mental retardation, epilepsy, ocular abnormalities, including coloboma, microphthalmos, cataracts, and skeletal, cardiac, and urogenital abnormalities. Associated abnormalities include hemihypertrophy, hemimegalencephaly, seizures, including infantile spasms, sensorineural deafness, spastic hemiparesis, kyphoscoliosis, and polydactyly. A tendency to malignant transformation of nevi and associated visceral malignancies (Wilms tumor, astrocytoma, intrathoracic teratoma) are reported. (Egan CA, et al. Neurologic variant of epidermal nevus syndrome with a facial lipoma. Int J Dermatol 2001 Mar;40(3):189-90).

CNS NEOPLASMS

CLINICO-RADIOLOGICAL PROFILE OF PEDIATRIC GLIOBLASTOMA

Researchers at Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India, studied the clinico-radiological profile, pathology, treatment and outcome of 65 pediatric patients (age < 18 years) with histopathologically proven diagnosis of intracranial glioblastoma. Male-to-female ratio was 2.6:1, with a mean age at diagnosis of 13.3 years (range 2-18 years). Most common presenting symptoms were headache with or without vomiting (n=51, 78%), seizures (n=42, 65%), and focal deficits (n=31, 47%). The tumor was supratentorial in 62 (95.4%) patients, frontal in 30%, and temporal in 9%; it was located deeply in 16 (25%), in the thalamus in 10 (12%). Obstructive hydrocephalus occurred in 13 (20%) patients and intratumoral bleeding in 5 (7.7%). Total tumor excision was achieved in 43 (66%) patients, and the remainder had incomplete excisions (n=22, 34%). Mean progression-free and overall survivals were 10 and 20 months, respectively; 3 patients survived for >5 years. Extent of resection was the independent predictor of survival (p=0.002). (Das KK, Mehrotra A, Nair AP, et al. Pediatric glioblastoma: clinico-radiological profile and factors affecting the outcome. Childs Nerv Syst 2012 Dec;28(12):2055-62). (Response: Dr Raj Kumar. E-mail: rajkumar1959@gmail.com).

COMMENT. Glioblastoma is an uncommon brain tumor in children compared to the prevalence in adults. Of all CNS tumors in children glioblastoma accounts for ~3–9%; the higher figure is that of Bailey P, Buchanan DN, and Bucy PC, in their classic study in Chicago (Bailey P, Buchanan DN, Bucy PC. Intracranial Tumors of Infancy and Childhood. Chicago: University of Chicago Press; 1939). The relative frequencies of the pathological varieties of intracranial space occupying lesions in children have changed over time; almost a century ago, tuberculoma was the most common lesion (Critchley M. Brain tumours in children: Their general symptomatology. Br J Child Dis 1925; 22:251-264).

The prevalence estimates for primary brain tumors in the United States are 35.4 per 100,000 person-years for children <20 years old and 278/100,000 for adults. (Porter KR, et al. Neuro Oncol 2010 Jun;12(6):520-7). The prevalence for malignant brain tumors in children is 25/100,000 and for non-malignant tumors, 11/100,000.