Sarcoma

A sarcoma (from the Greek σάρξ sarx meaning “flesh”) is a cancer that arises from transformed cells of mesenchymal origin. Thus, malignant tumors made of cancerous bone, cartilage, fat, muscle, vascular, or hematopoietic tissues are, by definition, considered sarcomas. This is in contrast to a malignant tumor originating from epithelial cells, which are termed carcinoma. Human sarcomas are quite rare. Common malignancies, such as breast, colon, and lung cancer, are almost always carcinoma.

Primary bone sarcomas

Soft tissue sarcomas

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see Gliosarcoma.

see Meningeal sarcoma.

see Rhabdomyosarcoma.

CIC-rearranged sarcoma.

Primary CNS sarcomas are very rare pediatric tumors with no defined standard of care.

A retrospective review of children diagnosed with a primary CNS sarcoma and treated at 2 Canadian tertiary care centers between 1995 and 2012.

Fourteen patients with nonmetastatic primary CNS sarcoma were identified; in 9 patients, tumors were located in the cerebral hemisphere and 7 of these patients presented with intratumoral hemorrhage. One infant who died of progressive disease postoperatively before receiving any adjuvant therapy was not included in this study. The final cohort therefore included 8 patients (4 males). Median patient age at diagnosis was 11.8 years (range 5.8-17 years). All tumors were located in the right hemisphere. Duration of symptoms prior to diagnosis was very short with a median of 2 days (range 3-7 days), except for 1 patient. Three (37.5%) patients had an underlying diagnosis of neurofibromatosis Type 1 (NF1). Gross-total resection was achieved in 5 patients. The dose of focal radiation therapy (RT) ranged between 54 Gy and 60 Gy. Concomitant etoposide was administered during RT. ICE (ifosfamide, carboplatin, etoposide) chemotherapy was administered prior to and after RT for a total of 6-8 cycles. Seven of the 8 patients were alive at a median time of 4.9 years (range 1.9-17.9 years) after treatment.

In this retrospective series, patients with primary CNS sarcomas located in the cerebral hemisphere most commonly presented with symptomatic acute intratumoral hemorrhage. Patients with NF1 were overrepresented. The combination of adjuvant ICE chemotherapy and focal RT provided encouraging outcomes [1)].

Spinal sarcoma

Spinal sarcoma

Synovial sarcoma

Synovial sarcoma

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Lafay-Cousin L, Lindzon G, Taylor MD, Hader W, Hawkins C, Nordal R, Laperriere N, Laughlin S, Bouffet E, Bartels U. Successful treatment of primary intracranial sarcoma with the ICE chemotherapy regimen and focal radiation in children. J Neurosurg Pediatr. 2016 Mar;17(3):298-302. doi: 10.3171/2015.6.PEDS14709. Epub 2015 Nov 20. PubMed PMID: 26588458.