Ewing’s sarcoma

Ewing’s sarcoma: aggressive malignant tumor with a peak incidence during the second decade of life.

Ewing's sarcoma is a primary bone tumor that affects mainly children and adolescents. It's one of a group of cancers known collectively as the Ewing sarcoma family of tumors – ESFT or sometimes just EFT. It's the second most common bone cancer in children, but it's also relatively uncommon. It accounts for only 1% of all childhood cancers. Although it can occur at any age, it very rarely occurs in adults over the age of 30.

Because many illnesses can cause the same symptoms as Ewing's sarcoma, it's sometimes missed in its early stages. But early diagnosis and treatment is important. If found early enough, before it spreads to multiple organs,

Vertebral column

The vertebral column is an infrequent site of primary involvement in Ewing sarcoma. Yet when Ewing sarcoma is found in the spine, the urge for decompression is high because of the often symptomatic compression of neural structures. It is unclear in alleviating a neurological deficit whether chemotherapy is preferred over decompressive laminectomy.

Primary intracranial Ewing sarcoma

Primary intracranial Ewing sarcoma.

Treatment

Treatment is mostly palliative: radical excision followed by RTX (very radiosensitive) and chemotherapy [1].

The use of stereotactic body radiotherapy (SBRT) is well-accepted [2].

SRS for spine metastases from Ewing sarcoma can be considered a treatment option in adolescent and young adult patients and is associated with acceptable toxicity rates. Further studies must be conducted to determine long-term local control and toxicity for this treatment modality [3].

Complications

SIADH.

Combined intracranial/extracranial lesions.

Spine metastases are more common than primary spine lesions.

Intramedullary Ewing's Sarcoma

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see Ewing's Sarcoma peripheral primitive neuroectodermal tumor

Case series

Fifteen patients with Ewing sarcoma primarily located in the spine were treated between 1983 and 2015. Localization, neurological deficit expressed as Frankel grade, and outcome expressed as Modified Rankin scale before and after initial chemotherapy, the recurrence rate, and overall survival were evaluated.

Nine patients (60%) were female. The age at presentation was 15.0 ± 5.5 years (range: 0.9-22.8 years). Ten patients (67%) were initially treated with chemotherapy, and 1 patient (7%) was treated primarily with radiotherapy followed by chemotherapy. The remaining 4 patients (27%) were initially treated with decompressive surgery. All patients treated primarily nonsurgically improved neurologically at follow-up, showing the importance of chemotherapy as an effective initial treatment option.

Adequate and quick decompression of neural structures with similar results can be achieved by chemotherapy and radiotherapy, avoiding the local spill of malignant cells [4].

Case reports

Intradural Extramedullary Ewing's Sarcoma in the Cervical Region [5].

A 21-year-old woman presenting with quadriplegia which was initially diagnosed with an epidural abscess in view of her MR scan and raised inflammatory marker levels. Histology revealed an epidural extra-osseous Ewing's sarcoma (EES). Epidural location of EES is a very rare condition which can be very challenging to diagnose. Early diagnosis and surgical excision followed by chemotherapy represent the main stem of management [6].

A case report demonstrates that hypofractionated partial-brain radiation therapy with limited margins is a reasonable approach following gross tumor resection of Ewing sarcoma metastases to the brain. The patient presented with 2 intracranial metastases treated with gross-total resection followed by radiation therapy to 30 Gy in 5 fractions. The patient experienced symptomatic treatment-related inflammatory changes with resolution after receiving dexamethasone. He remains alive at 21 months of follow-up with no evidence of disease [7].

References

1)
Grubb MR, Currier BL, Pritchard DJ, et al. Primary Ewing's Sarcoma of the Spine. Spine. 1994; 19:309–313
2)
Karimi AM, Campbell SR, Parsai S, Angelov L, Scott J, Qi P, Anderson P, Chao ST, Murphy ES. Aggressive Local Control With Multisite Stereotactic Body Radiation in Metastatic Ewing Sarcoma: A Literature Review and Case Report. Anticancer Res. 2020 Feb;40(2):951-955. doi: 10.21873/anticanres.14028. Review. PubMed PMID: 32014939.
3)
Parsai S, Juloori A, Angelov L, Scott JG, Krishnaney AA, Udo-Inyang I, Zhuang T, Qi P, Kolar M, Anderson P, Zahler S, Chao ST, Suh JH, Murphy ES. Spine radiosurgery in adolescents and young adults: early outcomes and toxicity in patients with metastatic Ewing sarcoma and osteosarcoma. J Neurosurg Spine. 2019 Nov 29:1-8. doi: 10.3171/2019.9.SPINE19377. [Epub ahead of print] PubMed PMID: 31783349.
4)
Mirzaei L, Kaal SE, Schreuder HW, Bartels RH. The Neurological Compromised Spine Due to Ewing Sarcoma. What First: Surgery or Chemotherapy? Therapy, Survival, and Neurological Outcome of 15 Cases With Primary Ewing Sarcoma of the Vertebral Column. Neurosurgery. 2015 Nov;77(5):718-25. doi: 10.1227/NEU.0000000000000903. PubMed PMID: 26308634.
5)
Warade AC, Jha AK, Pattankar S, Madiwale C, Rekhi B, Desai K. A Rare Case of Intradural Extramedullary Ewing's Sarcoma in the Cervical Region. Neurol India. 2019 Nov-Dec;67(6):1543-1544. doi: 10.4103/0028-3886.273622. PubMed PMID: 31857557.
6)
Bailey M, Mccabe M, Pal P, Agushi E, Karabatsou K. Cervical epidural extra-osseous Ewing sarcoma mimicking an epidural abscess. Br J Neurosurg. 2016 Feb;30(1):113-114. Epub 2015 Sep 7. PubMed PMID: 26982951.
7)
van Dams R, Park HS, Alomari AK, Ricciardi AS, Rao H, McNamara J, DiLuna ML, Bindra RS. Adjuvant hypofractionated partial-brain radiation therapy for pediatric Ewing sarcoma brain metastases: case report. J Neurosurg Pediatr. 2016 Apr;17(4):434-8. doi: 10.3171/2015.8.PEDS15313. Epub 2015 Dec 4. PubMed PMID: 26636250.