Primary intracranial Ewing sarcoma

- Primary Ewing Sarcoma of the Ethmoid Sinus With Intracranial Involvement: A Rare Case and Literature Review
- Challenges in Diagnosing Primary Intracranial Ewing Sarcoma/Peripheral Primitive Neuroectodermal Tumor: A Case Report
- Primary intracranial extraosseous Ewing's sarcoma with intraspinal metastasis in children: illustrative case
- Primary and metastatic cerebral Ewing's sarcoma: A case report about a rare entity and literature review
- Primary Intracranial Ewing Sarcoma With an Unusual Presentation: A Case Report
- Primary Intra-cranial Ewing's Sarcoma: An Exceptionally Rare Occurrence
- Primary intracranial peripheral primitive neuroectodermal tumor: lessons from an exceptionally rare neoplasm. Illustrative case
- Primary intramedullary extradural Ewing sarcoma

see Ewing's Sarcoma peripheral primitive neuroectodermal tumor.

EWS affecting the central nervous system (cranial and spinal column) is relatively rare with an annual incidence of approximately one case per million in the Western population. Due to their rarity, very few studies are available in the literature.

Kumarasamy et al. present the experience of managing 21 such cases, highlighting their clinical, and radiological findings, treatment strategy, and surgical outcomes in patients with primary EWS affecting the central nervous system.

They retrospectively collected hospital records of patients with primary EWS affecting the CNS (cranial and spinal column), who had been surgically treated in our Neuroscience Center between 2015 and 2023. Patients' demographics, presentation, radiological findings, treatment strategy including surgery and biopsy followed by adjuvant therapy, and outcome at discharge, and the latest follow-up were analyzed from our database.

There were sixteen male and five female patients with a mean age of 18.22 ± 12.73 years (ranging from 6 months to 59 years). The commonest presentation was headache and vomiting in cranial lesions (5/13 patients, 38.46%), and back pain in spinal lesions (4/8 patients, 50%). The site of lesions was cranial in thirteen patients (61.9%) and spinal column in eight patients (38.1%). The commonest site was the frontal region in the cranial group (6 patients, 46.15%) and the lumbar region in the spinal group (4 patients, 50%). All patients underwent surgical intervention [tumor resection (18) and biopsy (3)]. Tumor resection was achieved in 18 patients (85.71%). The extent of resection was gross-total excision in 9 patients (42.86%), near-total excision in 5 patients (23.8%), and tumor decompression in 4 patients (19%). Four patients underwent spinal instrumentation. Fifteen patients (71.42%) received multiagent chemo-radiotherapy according to institute protocol. Five patients (23.8%) with poor KPS expired within 6 months of surgery and could not receive adjuvant therapy. Two patients (9.52%) improved symptomatically, and nine patients (42.86%) remained asymptomatic at a mean follow-up of 25.1 ± 29 months. One patient (4.76%) after receiving 2 cycles of adjuvant therapy had initial improvement but expired later due to disease progression. Four patients after adjuvant chemo-radiotherapy (19%) expired due to systemic spread.

Primary EWS affecting the central nervous system is a rare variety. A detailed radiological assessment can aid in adequate planning for safe maximal resection. Timely tissue diagnosis is essential for initiating early treatment. Radical excision followed by adjuvant therapy offers a favorable outcome. Postoperative adjuvant chemo-radiotherapy aids in optimal disease control and surgical outcome. With the current treatment strategy, good surgical outcomes can be achieved. However, metastasis is not uncommon and should be evaluated regularly [1)].

Despite the inherent limitations of a retrospective, small cohort study, the work offers valuable insights into treatment strategies, surgical outcomes, and the role of adjuvant therapy. It underscores the need for careful pre-operative planning, radical excision, and post-operative chemo-radiotherapy to achieve favorable outcomes. However, further studies with larger sample sizes, longer follow-up periods, and more detailed data on metastasis and long-term outcomes are needed to refine treatment approaches and improve survival rates for this rare and challenging condition.

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Kumarasamy S, Garg K, Singh PK, Kumar A, Sharma R, Kedia S, Mishra S, Borkar S, Sawarkar DP, Verma SK, Gupta S, Satyarthee G, Garg A, Sharma MC, Kumar R, Singh M, Suri A, Chandra PS, Kale SS. Primary Ewing's Sarcoma affecting the Central Nervous System: A single-center experience and Narrative review. Childs Nerv Syst. 2024 Dec 13;41(1):46. doi: 10.1007/s00381-024-06707-y. PMID: 39671104.