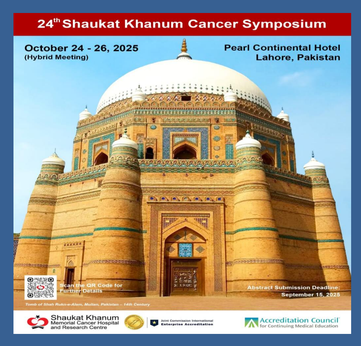
A Rare Case of Pediatric Plasmablastic Lymphoma With Widespread Skeletal Lytic Lesions: A Diagnostic Challenge

Dr. Novera Khalid · Dr. Saadia Anwar · Dr. Mehwish Faizan · Dr. Luqman Iqbal



Introduction

Plasmablastic lymphoma (PBL) is a rare and highly aggressive subtype of diffuse large B-cell lymphoma, usually associated with immunodeficiency such as HIV infection. It is primarily seen in adults, and pediatric cases are exceptionally uncommon

Case Presentation

An eight-year-old male presented with progressive right-sided cheek swelling, fever, and weight loss for last four months. One year back he was treated as degenerative brain disease because of difficulty in walking and MRI brain revealed nonspecific findings. On examination, there was a firm right cheek swelling without overlying skin changes. The rest of the systemic examination was unremarkable. His investigations revealed:

- **X-ray skull and skeletal survey:** Revealed multiple lytic lesions.

- **Pan CT scan:** Mass (6.0*7.4*6.2cm) involving right maxillary, ETHMOID sinus and nasal cavity causing distortion of right globe, extending up to right infra temporal fossa. Demonstrated variable-sized lytic lesions in both axial and appendicular bones with osseous and extra-osseous components, predominantly involving the calvarium and facial bones. CT chest normal.

- **Histopathology**: Initial biopsy of right maxilla raised the suspicion of metastatic Ewing sarcoma/RMS due to round blue cell tumor, further immunohistochemistry with positive CD138 and MUM1, confirmed the diagnosis of plasmablastic lymphoma with metastatic bone marrow infiltration.

- **HIV:** reactive

On the basis of above lab findings, antiretroviral started with 2 cycles of COP therapy given, after which there was reduction in tumor mass on reassessment CT but patient condition kept on deteriorating and could not survive.

Discussion

Pediatric plasmablastic lymphoma (PBL) is exceptionally rare, with fewer than 10 cases reported worldwide. The oral cavity is most common site of involvement with male predominance. Our case is unique due to widespread skeletal lytic lesions, a presentation seldom described in literature. Diagnosis is challenging and relies on immunohistochemistry showing CD138 and MUM1 positivity with loss of CD20, consistent with findings by Castillo et al. (2008, 2015). Despite therapy, PBL remains highly aggressive with poor outcomes.

Conclusions

Plasmablastic lymphoma should be suspected in children presenting with multiple lytic bone lesions especially in immunodeficent children. Early biopsy, detailed immunohistochemistry, and multidisciplinary approach is vital to accurate diagnosis of this rare disease.

Contact

Dr. Novera Khalid
University of Child Health sciences The Children Hospital Lahore
Department of Pediatric Hematology & Oncology
Email: Novera_khalid@yahoo.com

References

1. The biology and treatment of plasmablastic lymphoma by Jorge J. Castillo, Michele Bibas, Roberto N. Miranda
2. HIV-associated plasmablastic lymphoma: Lessons learned from 112 published cases by Jorge Castillo, * Liron Pantanowitz, and Bruce J. Dezube