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Case #9

Jesse Lee Kresak, Marie Rivera-Zengotita, Ahmad Alkhasawneh, Samer Al-Quran, Anthony T. Yachnis
University of Florida
Disclosures

• No financial relationships to disclose
Clinical History

- 64-year-old female presents with constant right-sided frontal headache radiating to cheek and jaw for one month.

- She has a past medical history of cervical cancer, myelofibrosis secondary to polycythemia vera, splenomegaly, and GERD.

- She receives blood transfusions approximately every 3 weeks

- She was hospitalized 3 weeks prior for urosepsis secondary to nephrolithiasis
• A left suboccipital craniectomy was performed
Diagnosis?
Flow Cytometry
Diagnosis

• Extramedullary involvement by chronic myeloproliferative neoplasm with extensive fibrosis
  • Consistent with prior bone marrow biopsy, 2011
  • Multilineage hematopoietic elements with erythroid and megakaryocytic hyperplasia with atypia and fibrosis
Chronic Myeloproliferative Neoplasm

- Polycythemia vera
- Essential thrombocytopenia
- Primary myelofibrosis
- Chronic myeloid leukemia
Polycythemia Vera

- Clonal proliferation of myeloid cells with the presence of an elevated red blood cell mass
- Occurs in all age groups, mean age 60
- Survival when treated exceeds 10 years, untreated 18 months
- 95 to 100% of patients with PV have a JAK2 mutation, usually V617F
Treatment

- Jakafi (ruxolitinib) is a JAK1/2 inhibitor
  - Improvement in debilitating symptoms and quality of life
  - Reduction in splenomegaly
  - ? Overall survival benefit
Secondary Myelofibrosis

- PV transforms into post-PV myelofibrosis in approximately 10% of patients
  - Risk factors: disease duration, age > 60
  - Non-clonal proliferation of fibroblasts caused by inappropriate secretion of growth factors by megas or platelets
- Significantly increases the risk for:
  - Acute leukemia
  - “Extramedullary hematopoiesis”
EMH Terminology

- Predominant sclerotic component and atypical megakaryocytes
- May be multiple or solitary
- Marker of advanced disease
EMH Tumor

• “Fibrous hematopoietic tumor”

• “Extramedullary myelofibrosis”
Extramedullary hematopoiesis involving the central nervous system and surrounding structures

Viktor Zherebitskiy MD, Carmen Morales MD, Marc R. Del Bigio MD, PhD

Human Pathology; 2011; 42 (10): 1524–1530

CASE OF MONTH MAY 2011

37-YEAR-OLD WOMAN WITH MULTIPLE INTRACRANIAL MASSES

Wen-Xia Jiang, MD; Shao-Qiang Zheng, MD; Pei-Jun Wang, MD

Brain Pathology; 2011, 21 (5): 607–610

CASE REPORT

Marie E. Beckner · John Y. K. Lee · Sydney S. Schochet Jr · Charleen T. Chu

Intracranial extramedullary hematopoiesis associated with pilocytic astrocytoma: a case report

Acta Neuropathology; 2003, 106: 584-587
Intracranial EMH - Treatment

• Radiation
  – Varying regimens 10-30 Gy
  – Good neurologic recovery

• Surgery - for decompression only

• Myelosuppression - already myelosuppressed
Patient Summary

• Low dose radiation
• Patient suffered a fall at home leading to left cerebellar hemorrhage
• Procedures were withheld due to thrombocytopenia
• Transferred to hospice and died one month later
Summary

- Patient with bilateral cerebellar masses
- History of myelofibrosis secondary to Jak2+ PV
- Dx: Extramedullary site of chronic myeloproliferative neoplasm
- Rare complication of hematopoietic dyscrasias
  - Thalassemia > myelofibrosis
- Metastatic vs reactive/reparative response
- Radiation alone