Coagulopathy was evident with prolonged PT (17.2 seconds) and elevated INR (1.46). Additional findings included suppressed TSH (0.07 mIU/L) suggesting hyperthyroidism and elevated ferritin (402 ng/mL). PET-CT performed on May 22, 2025, showed regression of previously enlarged cervical, axillary, iliac, and inguinal lymph nodes, indicating prior treatment response. However, a large hypermetabolic splenic lesion measuring 103 × 60 × 61 mm with SUVmax 32.82 was identified, with evidence of lateral capsular invasion. No bone marrow or hepatic FDG uptake was observed. Subsequent CT imaging on July 24, 2025, revealed alarming findings: intraparenchymal and subcapsular splenic hematoma with perihepatic, perisplenic, and pelvic free fluid consistent with hemoperitoneum. Additional incidental findings included a 17 mm right thyroid nodule and minimal left pleural effusion. Splenic tru-cut biopsy performed on July 17, 2025, confirmed diffuse large B-cell lymphoma with germinal center phenotype. Immunohistochemistry showed CD20(+), Bcl-2(+), Bcl-6 (+), MUM-1(+) with high proliferation index (Ki-67: 80%) and elevated c-Myc expression (60%). CD10 and CD5 were negative. The clinical constellation of findings confirmed Richter transformation with splenic DLBCL complicated by spontaneous splenic hemorrhage and hemoperitoneum, representing a medical emergency. **Discussion:** This case demonstrates an exceptionally rare presentation of Richter transformation. While most Richter transformations present with rapidly enlarging lymph nodes, isolated splenic involvement is uncommon. The extremely high SUVmax (32.82) indicated aggressive disease with high metabolic activity, consistent with high-grade DLBCL. The development of spontaneous splenic hematoma likely resulted from tumor infiltration weakening the splenic capsule and parenchyma, combined with underlying thrombocytopenia and coagulopathy. The resulting hemoperitoneum represents a life-threatening complication requiring urgent intervention. The coagulopathy and cytopenias observed may reflect both disease progression and splenic sequestration. The concurrent thyroid abnormalities warrant investigation for secondary malignancies or treatment-related complications. Management challenges include balancing the need for immediate treatment of aggressive lymphoma against the risk of exacerbating bleeding complications. Careful coordination between hematology, surgery, and radiology teams is essential for optimal outcomes. **Conclusion:** Richter transformation can present with rare but life-threatening complications including spontaneous splenic hemorrhage. High clinical suspicion, urgent imaging, and multidisciplinary management are crucial for patients with CLL developing new abdominal symptoms. This case underscores the importance of recognizing atypical presentations of Richter transformation to ensure prompt diagnosis and appropriate intervention.

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Adult Hematology Abstract Categories

Chronic Myeloproliferative Diseases

PP 23

CD56-Negative Conjunctival Solitary Extramedullary Plasmacytoma with Bence–Jones Lambda: Organ-Sparing Therapy and Durable Remission in a Young Adult

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Introduction: Solitary extramedullary plasmacytoma (EMP) accounts for a small fraction of plasma-cell neoplasms and rarely involves the conjunctiva. Distinguishing localized EMP from ocular adnexal lymphomas and reactive plasmacytosis is crucial, as management and prognosis differ substantially. We report a CD56-negative, lambda-restricted conjunctival EMP in a 27-year-old male with baseline Bence–Jones proteinuria, successfully treated with organ-sparing surgery plus orbital radiotherapy (RT) and maintained in remission at one year. **Methods:** This single-patient case review used prospectively recorded clinical data. Diagnostic workflow comprised ophthalmologic examination, complete blood count and chemistry, serum protein electrophoresis (SPEP) with immunofixation, 24-hour urine immunofixation, bone marrow aspirate/biopsy with immunohistochemistry (IHC), whole-body PET/CT, and brain/orbital MRI as indicated. Response was assessed clinically, biochemically (paraprotein clearance), and radiologically. **Results:** A painless, 1.5-cm vascular conjunctival mass was excised. Histology showed dense plasmacytic infiltration. IHC: CD38+, CD138+, lambda light-chain restriction, CD56−; B- and T-cell markers were non-diagnostic for lymphoma. SPEP showed no serum M-spike, while urine immunofixation revealed monoclonal lambda (Bence–Jones) positivity. Bone marrow morphology and flow cytometry demonstrated normal hematopoiesis without clonal plasma cells. PET/CT showed avid uptake confined to the conjunctival lesion; minor uptakes in stomach/sacrum lacked structural correlates. CRAB criteria were absent. Definitive local therapy consisted of adjuvant orbital RT (40 Gy in 20 fractions) after complete excision. Treatment was well tolerated. At 3 months, urine monoclonal lambda resolved; at 12 months, there was no local recurrence or new systemic disease clinically or on surveillance imaging/labs. **Discussion:** Key learning points include: (i) Localization and phenotype—conjunctival EMP is exceptional; CD56 negativity, while not universal, may be more frequent in extramedullary disease and can correlate with reduced bone tropism, supporting a truly localized process. (ii) Diagnostic clarity—lambda

restriction with CD38/CD138 positivity and absent marrow disease distinguishes EMP from ocular adnexal MALT lymphoma and IgG4-related disease. (iii) Therapeutic strategy—organ-sparing RT at 40–45 Gy achieves excellent control in EMP; here, 40 Gy sterilized the lesion and eliminated Bence–Jones proteinuria, implying the conjunctival clone was the source of the paraprotein. (iv) Surveillance—despite remission, EMP carries a risk of progression to multiple myeloma; our young patient remains on structured follow-up (periodic CBC, renal function, calcium, SPEP/IFE, serum free light chains, and symptom-directed imaging). **Conclusion:** This CD56-negative conjunctival EMP in a young adult underscores that meticulous staging can confirm true localization, enabling conservative surgery plus moderate-dose orbital RT to deliver durable biochemical and clinical remission. The rapid clearance of Bence–Jones lambda after RT highlights the curative potential of localized therapy while reinforcing the need for vigilant long-term monitoring.

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PP 24

TRANSFORMATION OF CHRONIC MYELOMONOCYTIC LEUKEMIA (CMML) INTO MYELOID SARCOMA: A RARE CASE WITH CERVICAL LYMPH NODE INVOLVEMENT

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Objective: Introduction: Chronic myelomonocytic leukemia (CMML) is a clonal hematologic malignancy with features of both myelodysplastic and myeloproliferative neoplasms [1]. Transformation into acute myeloid leukemia (AML) occurs in 15–20% of cases, while extramedullary presentation as myeloid sarcoma is exceedingly rare and associated with aggressive disease and poor prognosis [2,7]. **Case report: Case Presentation** A 64-year-old male diagnosed with CMML in 2024 was treated with azacitidine, achieving hematologic response after four cycles. Following the tenth cycle, he developed a cervical mass with compressive symptoms. Excisional biopsy confirmed myeloid sarcoma involving the cervical lymph node. Concurrent bone marrow analysis revealed 100% cellularity with grade 2/4 reticulin fibrosis, monocytic proliferation, and 15–16% blasts, consistent with CMML-2. Immunohistochemistry showed CD33+ and MPO+ staining, negative for CD34, CD117, and TdT. Systemic chemotherapy was planned, but the patient deteriorated rapidly with pneumosepsis and died. **Conclusion:** DiscussionExtramedullary transformation of CMML into myeloid sarcoma is a rare clinical event, with limited cases reported [3]. Diagnosis can be challenging due to morphologic overlap with lymphoma, underscoring the necessity of immunophenotypic confirmation [6]. Therapeutic options remain limited, ranging from AML-type induction regimens to hypomethylating agents combined with venetoclax, and allogeneic stem cell transplantation for eligible patients [4,5,8]. However, outcomes remain poor, with median survival after extramedullary progression of ~6 months [1,7].ConclusionThis case illustrates the rare transformation of CMML-2 into myeloid sarcoma with cervical lymph node involvement, highlighting diagnostic complexity, limited treatment options, and rapid disease progression. Early biopsy of new masses and bone marrow reassessment are crucial for timely diagnosis, while novel therapeutic strategies are urgently needed to improve outcomes.

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PP 25

Familial HFE Hemochromatosis in Two Siblings: Clinical Course and Response to Deferasirox

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Introduction: Hereditary hemochromatosis (HH) is an autosomal recessive disorder characterized by excessive intestinal absorption of iron and progressive iron overload. Clinical features may include hepatomegaly, cirrhosis, diabetes, cardiomyopathy, hypogonadism, and arthropathy. Diagnosis is based on transferrin saturation, serum ferritin, and confirmation by genetic testing. We present two siblings with homozygous HFE gene mutation who showed marked hyperferritinemia and a favorable response to oral deferasirox therapy. **Methods (Case Presentation):** A 40-year-old male presented to the hematology outpatient clinic on January 17, 2025, with elevated ferritin levels. His 48-year-old brother had been diagnosed with primary hereditary hemochromatosis in 2011. The elder sibling was treated with oral desferrioxamine (1 × 3 tablets daily) for several years but discontinued therapy in 2020 and remained untreated thereafter. Genetic analysis demonstrated that both siblings carried the HFE c.187C>G (p.His63Asp) homozygous mutation. At presentation, the proband's serum ferritin level was 1845 ng/mL, while his brother's level exceeded 1600 ng/mL. Both patients were started on oral deferasirox at a dose of 20 mg/kg/day (1 × 6 tablets). Regular laboratory follow-up was conducted every 4–6 weeks. **Results:** After initiation of deferasirox, both siblings demonstrated significant biochemical improvement. • The proband's ferritin decreased from 1845 ng/mL to 1090 ng/mL within three months. • The elder sibling's ferritin declined from >1600 ng/mL to 945 ng/mL in the same period. Both patients tolerated the medication well, without major adverse events. No hepatic decompensation, cardiac dysfunction, or endocrine complications were observed during follow-up. **Discussion:** This familial case illustrates several important points. First, family history and genetic testing remain critical tools in early recognition of hereditary hemochromatosis. The diagnosis in the younger sibling was established promptly because of the known history in his elder brother. Second, although phlebotomy remains the standard of care in HH, oral iron chelators such as deferasirox may be