

was made. We continued administering oral prednisolone and danazol without transfusion for 3 years. The need for platelet transfusion guided us to schedule HSCT. Given the absence of any matched family donor in her case, a 9/10 HLA matched donor was found from the national stem cell bank. Reduced-intensity conditioning regimen (Fludarabine, 30 mg/m²/day, days -7 to -3; cyclophosphamide; CY, 10 mg/kg/day, days -6 to -3) and serotherapy (ATG, 10 mg/kg/day, days -4 to -2) were performed. Mesenchymal stem cells (MSC) infusion (1×10^6 /kg) was administered on days -1 and +7, along with a dose of 6.2×10^6 /kg peripheral stem cells. Tacrolimus, methotrexate, and prednisolone (1 mg/kg/day, 28 days) were administered as graft versus host disease (GVHD) prophylaxis. Neutrophil engraftment ($2020/\text{mm}^3$) occurred on the 9th day, platelet engraftment ($135000/\text{mm}^3$) occurred on the 12th day. She had CMV reactivation in the 3rd month of HSCT. Antiviral treatment for CMV infection was carried out for 3 weeks. On day +100, a steroid was added due to grade II skin acute GVHD (aGVHD). Following its tapering off after 15 days, steroid administration was stopped. The patient achieved complete chimerism, allowing the discontinuation of immunosuppressive treatments in the first year itself. **Discussion:** MRD and MUD transplants yield the highest success rate in patients with FA. However, the results of HSCT from an alternative donor are still unsatisfactory. MSCs are responsible for immune regulation, tissue repair and regeneration, homing, and support of the hematopoietic system. It has been reported that infusion of MSCs can reduce the development of aGVHD by 3-fold and improve the OS of patients after allogeneic HSCT in comparison to standard prophylaxis. The addition of MSC to the conditioning regimens for MMUD transplants in patients with FA has been proven advantageous due to its graft-supporting, immunosuppressive, and immunomodulatory properties. However, large-scale randomised controlled trials are yet required to back these benefits.

Keywords: Fanconi anemia, HSCT, mesenchymal stem cell

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Adult Hematology Abstract Categories, Transfusion Medicine and Apheresis

PP 15

GRANULOCYTE TRANSFUSION ACCELERATES RECOVERY FROM NEUTROPENIA IN PATIENTS WITH HEMATOLOGIC MALIGNANCIES

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Objective: Neutropenia is the most common and serious consequence of myelosuppressive chemotherapy in patients with hematologic malignancies. Granulocyte transfusions can

restore granulocyte counts and thus theoretically reduce the risk of infection in such patients. In our study, we aimed to demonstrate the efficacy of granulocyte transfusion in neutropenic patients with hematologic malignancy despite recombinant myeloid growth factor therapy. **Methodology:** In this retrospective study, 72 patients who were treated in our hematology clinic between 2016 and 2022 and who met the criteria of our study were included. Demographic data, malignancy subtypes, chemotherapy regimens, number of neutropenic days, clinical outcome before and after granulocyte transfusion, and neutrophil count changes in blood parameters were analyzed. In the study, p-values less than 0.05 were considered significant. The analyses were analyzed with the SPSS 25.0 program. **Results:** In our study, 56.9% of the patients were male, the most common diagnosis was AML with 65.3% and 91.7% Gram-/ + was the most common type of treatment. It was observed that 62.5% of the patients recovered from neutropenia after granulocyte transfusion and 37.5% did not recover or exited. It was observed that patients who were neutropenic before chemotherapy were more likely to recover from neutropenia after granulocyte transfusion ($p=0.01$) and had lower rates of recovery from neutropenia ($p=0.04$). **Conclusion:** Considering the present results, granulocyte transfusion seems to accelerate the recovery from neutropenia in the sample we analyzed. In addition, the diagnosis of the patient, the type of chemotherapy received, and the time of granulocyte transfusion were evaluated as factors affecting the results. However, in light of the data obtained, we believe that prospective studies with a larger number of patients should be conducted to evaluate the consistency of our results.

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Adult Hematology Abstract Categories, Other Diseases

PP 16

CARCINOMA EX PLEOMORPHIC ADENOMA: DIAGNOSTIC CHALLENGE AND TREATMENT PROTOCOL

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Objective: Carcinoma ex pleomorphic adenoma CXPA, a rare epithelial malignancy arising from a primary or recurrent benign pleomorphic adenoma, accounts for 11.% of all malignant salivary gland neoplasms. It is difficult to diagnose pre-operatively. often poses a diagnostic challenge to clinicians and pathologists Treatment involves an ablative surgical procedure with neck dissection followed by radiotherapy. We aim to investigate the impact of postoperative radiotherapy on improving disease-free survival. **Case report:** A 39-year-old Libyan male presented with painless swelling near the angle of the right mandible four months ago. FNA Cytology showed a benign pleomorphic adenoma. A total parotidectomy with VII CN preservation was done in September 2022. The histopathological features were consistent with carcinoma EX pleomorphic adenoma, a widely invasive salivary duct

carcinoma grade III with < 1mm(close)margins staged PT1 N0 M0. The immunohistochemistry revealed the negative expression of ER and PR assays. **Methodology:** In December 2022, he received adjuvant radiation to the tumor bed (66 GY) in 33 fractions over 6 weeks based on the VMAT technique. 12-month follow-up, the patient showed no evidence of local or regional disease recurrence or distant metastasis. **Results:** Radical surgery, followed by adjuvant radiotherapy, should be considered the standard of care for a patient, with significant improvement in 5-year locoregional control. and in general, salivary gland neoplasms respond poorly to chemotherapy and are currently indicated only for palliative sitting. More prospective data is needed to establish a role for hormonal therapy and molecularly targeted therapies. **Conclusion:** CXPA is an uncommonly aggressive malignancy that, if associated with regional metastasis, invariably leads to mortality. Total resection of the tumor, followed by adjuvant radiotherapy, should be considered the standard of care for a patient with significantly improved 5-year locoregional control. Early and prompt diagnosis, followed by aggressive surgical intervention and adjuvant radiotherapy for patients with carcinoma ex pleomorphic adenoma, can enhance their survival rates.

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PP 17

AGGRESSIVE SALVAGE THERAPY OF OLFACTORY NEUROBLASTOMA CASE REPORT EXPERIENCE

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Objective: Olfactory neuroblastoma (ONB) is a rare malignant neoplasm arising from the olfactory neuroepithelium. It accounts for 3–5% of all nasal and Sinonasal malignancies, with an incidence of approximately 0.4 cases per million. A complete surgical resection of tumor followed by a full course of radiotherapy, is considered the treatment modality of choice for most ONBs. We aim to assess the impact of aggressive salvage radiotherapy in olfactory neuroblastoma on local recurrence and overall survival. **Case report:** A 41-year-old Libyan female presented in 2020 with a mass in the right nasal cavity that caused persistent nasal congestion with intermittent epistaxis over one year ago. Histopathological characteristics and immunohistochemical findings of the biopsy confirmed an olfactory neuroblastoma grade III, Radiological imaging evaluation revealed group B stage, and an incomplete excision was done, followed by radical radiotherapy with 70 GY in 35 fractions over 5 weeks to the residual disease. **Methodology:** Imaging follow-up for three years up to February 2024 shows no signs of local recurrence or distant metastasis. **Results:** Although multi-disciplinary care is required, surgical treatment alone is effective for low-grade tumors with free margins. Adjuvant radiation is used for low-grade tumors with close margins, residual disease, or recurrent disease, and for all high-grade cancers. The poor

prognosis associated with high-grade tumors may also mandate the addition of chemotherapy. Because recurrence can occur after 5 or even 10 years, aggressive management and long-term follow-up are mandatory. **Conclusion:** Multimodal therapy, including post-operative radiotherapy of high-grade incompletely resected ONB, with precise treatment planning based on CT simulation, could achieve an excellent local control rate with acceptable toxicity and reasonable overall survival for patients with ONB. Still, the rarity of the disease makes it difficult to draw definitive conclusions about the role of systemic treatment in induction and concomitant settings.

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PP 18

SEVERE CONGENITAL NEUTROPENIA WITH GLUCOSE-6-PHOSPHATASE CATALYTIC SUBUNIT 3 (G6PC3) DEFICIENCY OR DURSUN SYNDROME DIAGNOSED AT ADULTHOOD

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Case report: Severe congenital neutropenia is rare and usually diagnosed at childhood. G6PC3 deficiency emerge by mutation in glucose metabolism controlling genes as a syndromic variant. We here present a young adult case with unexplained neutropenia after kidney transplantation for FMF related AA amyloidosis. He had facial dysmorphism, growth retardation, and atrial septal defect. Parents were relatives and he had recurrent infection history. Genetic screening revealed G6PC3 gene mutation in patient.

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PP 19

THE RARITY OF PRIMARY CUTANEOUS MALIGNANT MELANOMA OF THE BREAST REQUIRES SPECIAL CONSIDERATION IN THE MANAGEMENT.

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Objective: Cutaneous malignant melanoma of the breast is a rare tumor, accounting for less than 5% of all malignant melanomas, Surgical resection is the commonly adopted treatment method for malignant melanoma, supplemented by chemotherapy, radiotherapy, and immunotherapy treatments, resulting in a comprehensive treatment strategy. We aim to assess the efficacy of adjuvant radiotherapy in managing cutaneous