resonance imaging (MRI) was the most widely used diagnostic modality (N=226). Overall, blood transfusion was the commonest reported treatment (N=30), followed by radiotherapy (N=20), surgery (N=15), hydroxyurea (N=12), steroids (N=6), and exchange transfusion (N=2). An outcome was reported in 20% of patients, all recovered, with no reported mortality. **Conclusion:** EMH is considered a rare phenomenon in BTM patients. It can occur in any organ system with varied clinical features. MRI can effectively diagnose EMH, and conservative management has similar results compared to invasive treatments. Larger studies, focusing on outcomes may enhance guidelines on preventive and therapeutic strategies for managing EMH in BTM.

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LATE DIAGNOSIS OF CONGENITAL METHEMOGLOBINEMIA IN A 33-YEAR-OLD CYANOTIC PATIENT, CASE REPORT AND REVIEW OF LITERATURE

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Objective: Due to mild symptoms, congenital methemoglobinemia is rarely diagnosed and reported as a cause of the cyanosis, especially in adults. Despite the benign nature of congenital methemoglobinemia, it is crucial to keep it in the differential diagnosis list when assessing cyanotic patients, mainly if he has a normal PaO2. Patients are usually asymptomatic and are treated for cosmetic purposes, but they might suffer from severe complications if exposed to oxidative agents. **Case report:** A 33-year-old lady presented to ED with difficulty breathing and bluish discoloration gradually increased over days, without fever or cough. she mentioned having recurrent similar episodes since childhood. family history positive for similar episodes in the sister. physical examination positive for central and peripheral cyanosis, with o2sat of 88% and RR of 20, the rest of examination within normal limits. laboratory tests normal except for ht 48.9%, PO2 160 on ABGs. **Methodology:** The patient's clinical picture of cyanosis with no evidence of cardiovascular or pulmonary diseases and the discrepancy between PaO2 and O2 saturation on oximeter required thinking of methemoglobinemia as a possible diagnosis despite the patient's age and the absence of any exposures. Methemoglobin level 20.9% (0-1.5%). Hemoglobin electrophoresis did not detect any abnormal hemoglobin. The activity of NADH cytochrome b5 reductase or the level were not done. **Results:** We searched PubMed and Google Scholar, we found 22 articles with a total of 30 patients with congenital methemoglobinemia. The mean age of the patients was 25 years (range 2 days-61 years); most of them were previously healthy. Out of 30 patients, 16 were treated with ascorbic acid or methylene blue or both with improvement, 14 either were not treated or treatment not mentioned in the report. Our patient received ascorbic acid 500 mg BID orally and improved clinically and laboratory. **Conclusion:** Due to mild symptoms, congenital methemoglobinemia is rarely diagnosed and reported as a cause of the cyanosis, especially in adults. Despite the benign nature of congenital methemoglobinemia, it is crucial to keep it in the differential diagnosis list when assessing cyanotic patients, mainly if he has a normal PaO2. Patients are usually asymptomatic and are treated for cosmetic purposes, but they might suffer from severe complications if exposed to oxidative agents.

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PP 50

A RARE ASSOCIATION IN A CASE WITH HEREDITARY SPHEROCYTOSIS: SPECTRIN BETA (SPTB) AND JAK-2 MUTATION

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Case report: Five types of gene variants are seen in hereditary spherocytosis (ANK, SPTB, SPTA1, SLC4A1, EPB42). JAK2 V617F mutation; is most common seen in bcr-abl negative chronic myeloproliferative diseases. As a result of NGS performed before splenectomy, SPTB c.4973+2T> C and JAK-2 c.1849G>T p.(Val617Phe) mutations were detected. Co-occurrence of these two mutations requires special attention in terms of the management of thrombocytosis and side effects that may occur after splenectomy.

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RETROSPECTIVE EVALUATION OF PATIENTS WITH LANGERHANS CELL HISTIOCYTOSIS FOLLOWED IN OUR CLINIC

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Objective: Langerhans cell histiocytosis (LCH) is a rare histiocytic disorder that can be especially seen in children and young adults. The clinical presentation of patients with LCH varies according to the sites of involvement. In about half of patients, the disease is limited to a single organ system and bone involvement is very common. In this study, it was aimed

to retrospectively evaluation the patients diagnosed with LCH who were followed up and treated in our clinic. **Methodology:** The data of patients over the age of 18 years who were followed up and treated in Bozyaka Training and Research Hospital Hematology Clinic between 2015-2021 were scanned retrospectively from the hospital system. **Results:** Data of 6 patients were obtained. The mean age of the patients was 33.6. There was no difference between the genders. Pain was the reason for admission in 4 patients and was the most common symptom. While the most frequently involved system was the skeletal system with 5 patients, lung involvement was seen in 2 patients. Vinblastine and prednisolone combination therapy was given to 1 patient, who developed steroid-related avascular necrosis. One patient who was planned for combination treatment **Conclusion:** LCH is a rare disease especially seen in children and young adults. It can involve the skeletal system, lungs, and other organs. The prognosis is often good with excision of the lesion or systemic treatment.

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PP 52

PILOMATRIX CARCINOMA WAS BEYOND ANY EXPECTATIONS: A CASE REPORT OF CARCINOMA CLINICIAN SHOULD BE AWARE OF IT

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Introduction: Pilomatrix carcinoma is a rare cutaneous tumor derived from follicular matrix with about 150 reported cases and it is considered as locally aggressive tumor with a tendency to recur. In addition to lymph node metastases and poor prognosis, it is non-chemotherapy responding in systemic metastasis. **Method and result:** A 37-year-old Libyan man presented with two large, coalesced nodules in the face measuring about 3*2 cm and 3*3 cm, treated as a case of lipoma by surgical excision based on clinical diagnosis, reappearing of larger nodule 9*6*4 cm involving almost all the left check, surgical excision was done with histopathological features of pilomatrix carcinoma infiltrating to the subcutaneous adipose tissue and deep striated muscle with no clear margins. Conventional radiotherapy by electron beam was started using the linear accelerating machine, with total radiotherapy dose 60 gray (Gy) in 30 fraction for six weeks. No local recurrences, nor lymph node or systemic metastasis since June 2020. **Conclusion:** Pilomatrix carcinoma must always be considered in the differential diagnosis of nodular tumors of the head-and-neck due to high recurrence rate after simple excision. Furthermore, local radiotherapy post incomplete surgical excision is the best adjuvant therapy.

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WHAT HAVE WE EXPERIENCED WITH COVID-19 IN PATIENTS WITH HEMATOLOGICAL DISORDERS?

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Objective: Patients with cancer are considered highly vulnerable to the COVID-19 disease. However, there are still few data in hematologic patients. Some small studies have shown a high mortality on patients with hematologic malignancies and COVID-19. In this study we aim to report a single center experience of the hematologic patient population with COVID-19 disease. **Methodology:** This single centre, retrospective, cohort study included a total of 111 adult patients (aged ≥18 years) with diagnosis of neoplastic and non-neoplastic hematologic diseases between March 2020 and August 2021. Ethics committee approval was obtained from the Istanbul University Istanbul Faculty of Medicine Clinical Research Ethics Committee. Categorical variables were compared using Pearson's Chi-square test. STATA16-MP was used for the statistical analysis. **Results:** A total of 111 patients (median age:55) with hematologic disease were diagnosed with COVID19. Ninety patients had neoplastic hematologic disorder. Fourty-five patients were receiving anti-neoplastic treatment at the time of COVID19 diagnosis. A total of 21 patients (overall mortality rate:19%) died and 19 of them had neoplastic disorder. The malignancy mortality rate was estimated to be 21%. 45 of 90 cases were receiving chemotherapy. Ten of these 45 patients (22%) died due to COVID19 disease. **Conclusion:** In our study the majority of patients who died due to COVID-19 had hematological malignancies. The cytokine storm which affects the clinical outcome in COVID-19 may contribute to dismal prognosis in hematologic malignancies in which cytokine increase is a part of process. Most of the succumbed patients were relapsed refractory multiple line treated which may reflect the immune insufficiency. It seemed COVID-19 progress is mostly poor in hematologic malignancies compared otherwise healthy people.

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A RARE CAUSE OF ANEMIA IN ADULTHOOD CONGENITAL DYSERYTHROPETIC ANEMIA

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