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.

https://doi.org/10.1016/j.htct.2021.10.1058

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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.

https://doi.org/10.1016/j.htct.2021.10.1059

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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.

https://doi.org/10.1016/j.htct.2021.10.1060

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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