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Case Report

Sweet Syndrome Leading to Diagnosis of Myelodysplastic Syndrome during Pregnancy

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Background

Myelodysplastic syndromes (MDS) are defined as a series of malignant hematological conditions in which hematopoiesis is dysplastic and ineffective. MDS have the potential to progress to Acute Myeloid Leukemia (AML), which is often refractory to treatment. MDS causes chronic cytopenias, such as neutropenia, thrombocytopenia and anemia. Patients affected by MDS are often asymptomatic, however, some may present with infection or symptoms of chronic anemia. Cutaneous manifestations, such as Sweet Syndrome and Myeloid Sarcoma are rare, and may herald a transition to AML. In the pregnant patient, many overlapping symptoms may be seen, including fatigue, anemia and immunocompromise. When determining delivery planning in the affected pregnant patient, the risks of neonatal prematurity should be weighed against the risk of delaying maternal treatment and transformation to AML.

Case Description

Patient is a 24 year old G2P1001 at 28 weeks gestation who initially presented to her PCP with complaint of new onset blistering rash on her hands and feet. She was treated with PO antibiotics, and topical steroid and antifungal creams without improvement. She was therefore referred to Dermatology. At the time of her referral visit, the rash was noticed to be spreading from hands/feet to elbows, ankles and knees. A punch biopsy was performed that revealed deep perivascular lymphohistiocytic infiltrate, neutrophils, and interstitial dermal mucin. A CBC was performed and found 7% blasts in circulation. Therefore, the rash was thought to be Sweet Syndrome, with an underlying systemic hematologic process. The patient was referred to Hematology/Oncology, and a bone marrow biopsy confirmed high grade myeloid neoplasm with high risk features, specifically chronic myelomonocytic leukemia (CMML-2) and refractory anemia with excess blasts (RAEB-2). A multidisciplinary conference was held at the time of diagnosis when the patient was 32w0d with Hematology/Oncology, Maternal Fetal Medicine, and Neonatology. A treatment plan was developed, and a scheduled induction of labor (IOL) was planned at 34w0d to allow for expedited treatment of maternal MDS. If any evidence of disease progression were to occur, the plan was for immediate delivery. She underwent a normal spontaneous vaginal delivery after IOL of a 1745 g (3 lb 13 oz) baby girl with APGARs of 8 and 9 at 1 and 5 minutes, respectively. At seventeen days postpartum, she began her chemotherapeutic regimen of Azacitidine and Lenolidomide.

Discussion

The estimated incidence of MDS is 4.1/100,000 in the United States, with the median age at diagnosis of 65 years. Age appears to be an independent risk factor for the development of disease, with an incidence of 89 per 100,000 for those aged >80. Although the pathogenesis of MDS is unclear, it is thought, that like most malignancies, a series of oncogenic mutations develop that result in malignant process. These oncogenic mutations may result de novo, or as a consequence of exposure to prior chemotherapy (specifically alkylating agents), radiation, or environmental exposure to benzenes. Benzenes can be found in high concentrations in cigarette and second hand smoke, oil refineries and petroleum based fumes, rubber manufacturers, and chemical or plastic manufacturing plants. Autoimmune connective tissue disorders have also been associated with diagnosis of MDS, including Sjogren's syndrome, polyarteritis nodosa, polymyalgia rheumatic, Behçet's syndrome, inflammatory bowel disease and pyoderma gangrenosum, but a causal link between these disorders and subsequent MDS has not been established.

Most patient's diagnosis of MDS comes as a result of abnormalities found routine laboratory draws prompting further evaluation. If symptoms are present, they are often attributed to resultant cytopenias. Anemia is the most common, and presents as fatigue, shortness of breath, tachycardia, and pallor. Physical findings of underlying malignancy are rare. Cutaneous manifestations, such as Sweet Syndrome or Myeloid sarcoma, are uncommon and typically represent transformation from MDS to AML. Diagnosis of MDS following abnormal blood count is confirmed by a large blast burden and dysplastic cells seen on bone marrow biopsy.

In the pregnant population, non-specific symptoms of anemia are heralded as gestation related. Dilutional anemia of pregnancy is the most common cause for anemia in the pregnant patient, followed closely by iron deficiency. Only 25 cases of MDS diagnosed during pregnancy have been reported. It is anticipated, that as the survival rate of childhood cancers increases, there will be an uptick in reproductive aged women diagnosed with MDS secondary to prior chemotherapy

exposure. Due to rarity of the diagnosis, evidence based treatment algorithms in this population are lacking. In addition, antineoplastic agents, such as Lenalidomide (a thalidomine analog) and azacitidine, are contraindicated during pregnancy.

Prognosis of MDS is based on multiple factors. The Revised International Prognosis Scoring System (IPSS-R) in MDS can be used as a resource to determine mean survival. The IPSS-R takes into account the percent of blasts in the bone marrow, genetic alterations, and presence and severity of cytopenias. Based on this calculator, our patient's score is 6.5 which confers a "high" risk disease with median time to transformation of AML of 1.4 years and median survival of 1.6 years from time of diagnosis.

Treatment of MDS is indicated in any patient with symptomatic cytopenias. For high risk disease, aggressive chemotherapy with Azacitidine or Decitabine followed by allogenic hematopoietic cell transplant (HCT) can be offered. Pretreatment with intense chemotherapeutic regimens is recommended to decrease bone marrow blast burden to <5% at the time of bone marrow transplant. HCT is the only treatment available with a potential for a cure. Supportive therapy with transfusions and antibiotic treatment is necessary for improved survival. Complete remission of the disease is the goal of therapy and is defined as bone marrow with <5% blasts and normal maturation of all cell lines.

Conclusions

Diagnosis and treatment of maternal conditions becomes complicated when a viable pregnancy is involved. In the case of maternal malignancies, the initiation of chemotherapeutic regimens must be weighed against the potentially lifelong complications of neonatal prematurity. In cases of maternal malignancy diagnosed at early gestational ages, the ethical controversy of pregnancy termination versus continued gestation remains. In this case, the diagnosis of maternal MDS occurred during the third trimester after viability, allowing for administration of antenatal corticosteroids and sufficient fetal development in utero prior to scheduled induction of labor. In women with high risk malignancies, delivery and treatment is often recommend, but the burden of the decision rests on the shoulders of the mother, as she is forced to tally the costs of both physical and emotional ramifications of her choice on herself, her family, and the unborn child.

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