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Successful Prophylactic Pulmonary Arterial Aneurysm Repair in Pregnant Women with Behcet's Disease

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Abstract: ***Behcet's Disease is a multisystem vasculitis characterized by recurrent aphthous ulcers, genital sores and uveitis. It runs a chronic course with unpredictable exacerbations and relapses which may be caused by an autoimmune reaction. A 24 year old pregnant women presented with a decreased visual acuity of the right eye, hemoptysis, diarrhea, and cough. On history and examination, she was revealed to have anterior uveitis, monoarticular arthritis, oral ulcerations, pulmonary artery aneurysm and erythema marginatum. The diagnosis was difficult given the wide array of symptoms, eventually, she was diagnosed with Behcet's disease. Symptomatic treatment was initiated, she was started on colchicine, and surgery was planned for the pulmonary artery aneurysm as symptoms worsened. The patient was followed up and successfully delivered a viable healthy male fetus.***

1. Background

Behcet's disease is important as it is a rare case with only around 1 in 100,000 cases being present in the western world and India, however being more common in the Middle East and Japan.[1] The case had the added factor of the patient being pregnant which is interesting as in most instances pregnancy usually causes remission of Behcet's however, in this case, there was a flare-up of symptoms. The importance of this report on Behcet's is to understand the diagnostic criteria and the various differentials in order to be able to treat patients better, with complaints similar to this case.

2. Case Presentation

An 24-year-old woman visited the hospital for the first time with chief complaints of diminished vision in the right eye, a productive cough, and diarrhea. She has a 2-year history of recurrent painful oral and genital ulcerations for which she did not take any medication. She attributed the ulcerations to her poor dental hygiene. The patient denied leaving the country or any contact with sick family members. On asking again she admitted on having a rash on her calf area which was tender and recurrent but relieved over time without any treatment. She complained of having pain in her left knee while walking long distances which had negatively impacted her ability to make a living. She did not take any medications, occasionally used paracetamol for a headache and body pains. Smoking history was positive(1 pack of cigarette/day) and stopped 1 months ago, occasional alcoholic. She was sexually active and had been married for 6 years, did not recall her last menstrual period. She did not know her family history but stated her father had passed away due to an unknown illness.

On examination, the patient was slender, alert, awake, oriented, well built and moderately nourished. Her generalized appearance was toxic, diaphoretic and skin was pale in color. Lead a stressful lifestyle, had easily fatigability and infrequent menses. Her blood pressure was 130/80 mmHg with a heart rate of 97 beats per minute. On general physical examination, there was decreased breath sounds on the right side with other systems being normal. She complained of diarrhea during her hospital stay and had generalized discomfort.

Examination of the oral cavity revealed multiple ulcerations, which measured around 0.5- 1.4 cm in diameter. They were present under the lips, on the inside of both cheeks and a few on the tongue. Some ulcers bled on touch and presence of an erythematous halo on parts of the tongue. The decision to do a biopsy was taken which on microscopic examination relieved the ulcer to have the mucosa of parakeratinized stratified epithelium with signs of hyperplasia. The surrounding tissue showed intense neutrophilic inflammatory infiltrate. Necrosis could also be seen at parts but no features of malignancy were seen on microscopy. The resultant diagnosis of the ulcerations was that of large and small aphthous.

The patient was referred to ophthalmology for the vision changes in the right eye. An exam showed there was diffuse redness of the eye and on further questioning, the patient revealed she saw objects floating in her line of site at various points of the day. She has a visual acuity of 14/20 in the right eye and 16/20 in the left eye, with color vision on Ishihara plates being normal. Fundus exam disclosed vitritis and optic disc swelling with an absence of venous pulsations. The anterior segment had the presence of a hypopyon. This lead to the diagnosis of anterior uveitis with a hypopyon.

The patient's knees were remarkably inflamed with patellar effusion. She had difficulty in walking and bending. X-ray showed arthritic changes on the left knee. The diagnosis of monoarticular arthritis was made after synovial fluid analysis had revealed inflammatory cells. The culture, however, was negative for any organisms. The characteristic appearance of the rash on her left calf area was of erythema marginatum which was confirmed by dermatology on

histology and was given corticosteroid cream to relieve the symptoms.

The patient's condition did improve with treatment and after prophylactic surgery was undertaken for her pulmonary arterial aneurysm. She was discharged on Colchicine 1.5mg, Triamcinolone acetonide as cream 0.1% for her oral ulcerations and erythema marginatum, and scopolamine 0.2% for her uveitis. The patient was advised to follow up every two weeks to keep her condition in check.

3. Investigations

A chest of the patient showed a 5x3cm opacification in the inferior lobe of the right lung along with pleural effusion. CT scan of the thoracic cavity with contrast revealed a large dilation on the inferior lobe, partially extending up to the hilum of the lung with adhesions to the chest wall. Confirmed by radiology the scan gave definitive proof that a pulmonary arterial aneurysm was present which amassed the lower vessels. A digital angiography was done to further confirm the diagnosis.

The patient was subjected to a full workup including a USG abdomen, Chest- CT scan, ESR, C-Reactive Protein, and routine labs. CBC showed an elevated white blood cell count. ESR was 40 mm/h and hematocrit was 30.1% with all other labs being normal. She was anemic with a hemoglobin of 8.2 gm/dL. Her ultrasound came back positive for pregnancy with a dating of one month.

4. Differential Diagnosis

The diagnosis and the treatment of the disease is one that is multidisciplinary in nature because of the various symptoms that plague the body. The main goal of the treatment is for the disappearance of the lesions, in order to keep the disease under stable condition, to prevent blindness and to prevent death in severe cases. Throughout the course of the interaction with the patient many different diagnoses occurred before coming to the final conclusion, with the first being that all of the symptoms were unrelated and the cause of the problems was tuberculosis due to the hemoptysis and the productive cough being 2 out of the 3 major complaints she came to the hospital with and the other symptoms being present beforehand but not treated. This theory was discarded when a sputum test was performed and that it came back negative 3 times which indicated tuberculosis was not the cause.

When the patient was referred to an ophthalmologist for the blurring of vision, the report returned with of anterior uveitis with a hypopyon which led to the belief that the cause of the disease was sarcoidosis due to the fact that the patient was also suffering from joint pains and rashes on parts of the body. This was however ruled out when the examination of the oral cavity showed numerous ulcerations not consistent with stress or a vitamin deficiency. The patient also did not present with a dry cough which is a key feature in sarcoidosis.

Due to canceling out all of the various differential diagnosis, finally, the conclusion was that the patient was suffering from Behcet's disease after extensive literature research with there being no universally accepted definition nor there being definitive correct diagnostic criteria, the main way to diagnose the disease is through ruling out other diseases and by using the International Study Groups criteria.[2] In the current case after ruling out Tuberculosis, Sarcoidosis and Crohn's disease, and with careful consideration of the oral ulcerations, uveitis and the recurrent joint pain the conclusion was made that the patient was suffering from Behcet’s disease. The key differential feature, in this case, was that the patient was pregnant along with a pulmonary arterial embolism present which is a rare complication and genital ulcerations were not present.

5. Treatment

The patient was given colchicine 1.5mg/day after the diagnosis of Behcet's was made, along with Triamcinolone acetonide as cream 0.1% in Orabase for her oral ulcerations. For the anterior uveitis scopolamine, 0.2% was given. After reviewing the literature on Behcet's she was started on benzylpenicillin as well. Treatment initially was thought to be working, but she was suffering from refractory arthritis, however, because she was pregnant we could not start her on azathioprine as it is teratogenic.[3]

Prophylactic surgery was done for a large aneurysm, it was excised and replaced with a dacron graft of 25 mm. Postoperatively she remained stable and the fetus was not affected.

6. Outcome and Follow-Up

The surgery has no complications, and the patient was living a worthwhile life. She continued to work and go about her daily life. The patient followed up with her symptoms in check. In the 37th week of her pregnancy, she experienced labor pains and was decided to take up a cesarean section to avoid any other vascular complications as she was already predisposed. A healthy viable male fetus was delivered. The patient is still continuously following up with the care of the hospital on her regime of colchicine, benzyl penicillin, Triamcinolone acetonide, and scopolamine.

7. Discussion

The disease of the silk road involves a multitude of systems and as each case has different presenting factors, diagnosis of the disease is difficult. Increased awareness of the International Study Group of Behcet’s disease criteria should be made, as it gives the required elements to make a diagnosis possible. Behcet’s disease is one that can vary dramatically in terms of its symptoms and morbidity, with severe manifestations ranging from blindness and to even death due to complications of systemic manifestations.[4]

The diagnosis and the treatment of the disease is one that is multidisciplinary in nature because of the various symptoms that plague the body. The main goal of the treatment is for the disappearance of the lesions, in order to keep the disease under stable condition, to prevent blindness and to prevent death in severe cases.[5] Throughout the course of treatment,

many different diagnoses occurred before coming to the final conclusion, including tuberculosis, sarcoidosis, Crohn's disease, and systemic lupus erythematosus.

The reason a prophylactic excision of the pulmonary artery aneurysm was done is due to the large size of aneurysm present. Pregnancy is documented to have higher rates of vascular complications as seen in a study done by Iskender C et al.[6] If the surgery was not taken up the then the likelihood of complications of an aneurysm such as rupture would have been much higher.

Behcet's disease has many unusual presentations and is very difficult to diagnose. This case had many interesting features which make it unique as the patient was pregnant thus causing a flare up and had a pulmonary arterial aneurysm which isn't well documented. The literature on the effects of Behcet's on pregnancy is mixed. A study published by Gungor et all states in their conclusion that pregnant women who do suffer from Behcet's have a high rate of vascular complications and miscarriages.[7] However, a study published by Noel N et al states that pregnancy was not affected by Behcet's and complications were unlikely and colchicine will improve the disease course in Behcets.[8]

On further research, it was found that pregnancy can cause flare-ups on Behcet's in some patients which would explain the onset of most of her symptoms in such a manner. This however only occurred in 8 % of the patients in a study of 26 women with 55 pregnancies but the number of studies is limited. The norm is that Behcet's will cause for a remission of symptoms but in this case, there was a flare-up.[9] The patient was consulted about the pregnancy and about the increased chance of complications due to her condition especially as there are active symptoms present. The likelihood of preterm delivery and low birth weight babies is increased due to Behcet's being active. The concern of treatment interfering with the pregnancy was taken into consideration, however, there is no evidence the colchicine has any teratogenic effects.[10]

Behcet's disease is a disease that is not well known to most doctors as the number of cases are rare and not well documented. As more diagnosis are being made, and as knowledge has increased about the disease more case studies and research are being conducted. The importance of ruling out differentials is very important in order to make a diagnosis in accordance with the International Guidelines for Behcet's disease. Thereby there should be an increased awareness of Behcet's and should be a differential in any case with an oral and/or genital ulcerations, uveitis, vasculitis and erythematous changes. The disease prognosis and range of symptoms vary from person to person. Studies are now being conducted in order to find a better indicator for the diagnosis of Behcet's disease and for better treatment protocols to prevent relapses of the disease.[11]

8. Learning Points/Take Home Messages

1) A high level of suspicion should be held when a patient presents with recurrent oral ulcerations with other symptoms suggestive of Behcet's
2) Pregnancy can cause a variety of symptoms of Behcet's even though this is not well documented
3) Need to terminate a pregnancy in cases of Behcet's is not indicated.
4) There should be increased awareness and research on Behcet's disease.

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