Pregnancy with Polyostotic Fibrous Dysplasia: A case report.

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ABSTRACT

We are reporting a very rare case of a woman diagnosed with polyostotic fibrous dysplasia who became pregnant without a history of infertility. Unexpectedly, her symptoms of fibrous dysplasia had improved during pregnancy. She was preferentially delivered by cesarean section without morbidities. The aim of reporting this case is to view the implications of pregnancy on the disease. On the other hand, there is limited and undetermined data in the literature on skeletal complications during pregnancy.

Keywords: Pregnancy, Fibrous dysplasia, Abnormal uterine bleeding, Infertility.

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Introduction

Fibrous dysplasia is a rare bone disorder (7%) where bones are replaced by abnormal fibrous connective tissue which weakens the bones, making it painful and prone to fractures. Etiology is unknown but some link it to gene mutation. It is usually diagnosed in young age with misshapen bones, but mild cases may go undiagnosed until adulthood. It may affect only one bone (monostotic disease) or multiple bones (polyostotic disease). It can affect any part of the skeleton, but the most affected bones are the long bones of the legs, bones of the face, the skull and the ribs. Management may require medical treatment as well as surgical intervention (1). Polyostotic fibrous dysplasia is a synonym for McCune-Albright syndrome which is a fibrous dysplasia that is accompanied with hormonal abnormalities and skin pigmentation (café au lait spots) which is an extremely rare disorder with estimated prevalence between 1/100,000 and 1/1,000,000 (2). Malignant transformation can happen in less than 1% (3).

Effects of pregnancy on skeletal outcomes have not been determined yet. They are limited to case reports of skeletal complications during pregnancy. This has led some to speculate that pregnancy increases polyostotic fibrous dysplasia activity, placing women at risk for poor skeletal outcomes (4, 5). Nevertheless, two cases of polyostotic fibrous dysplasia in pregnancy have been reported. They were delivered vaginally without any reported complications (6).

Case report

A thirty years old woman, a known case of polyostotic fibrous dysplasia attended our antenatal clinic at King Hussein Medical Center on the 1st of June 2019 at 38 weeks gestational age for planning delivery. She was asymptomatic, with good fetal movements, reactive cardiotocography and obstetric ultrasound examination goes with gestational age.

She was booked for a general obstetrician and gynecologist in the private sector with regular antenatal follow ups and was referred to us because she has military health insurance. Her pregnancy went smoothly apart from threatened miscarriage. The patient has declared that her generalized bone pain and fatigue that she used to have due to her disease had improved as she got pregnant, and no complications of her disease was encountered ante partum.

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She was G2P0+1 and her first pregnancy ended with first trimester spontaneous complete miscarriage
Her menarche started at 15 years of age. She had irregular menstruation with history of months of amenorrhea which was treated by private sector with oral combined contraceptive drug. According to that, she had regular menstruation. She also had physiological dysmenorrhea which was relieved by paracetamol. She used combined oral contraceptive for three years after she got married and after stopping it, she got pregnant instantly without any history of infertility.
The patient was diagnosed with polyostotic fibrous dysplasia in 2012 at King Hussein Medical Center. It was affecting multiple bones including the skull, the thoracic and lumbar vertebrae and the sacrum. Brain computed tomography showed a huge bone hyperostosis with ground glass appearance, representing fibrous dysplasia in the left side of her face, left maxilla, left frontal bone and left nostril reaching supra sellar and infra sellar, pituitary extension could not be evaluated because of the huge lesion compressing brain tissue. Spine magnetic resonance imaging showed multiple varying sizes cystic changes in the vertebrae bodies, mainly at T10, L2, L5 and the sacral bone, a collapsed body of L5 with herniated disc into the end plate, and lumbar scoliosis convexity towards the left. She had been seen by a multidisciplinary team of orthopedics and endocrinologists and she was prescribed Somatostatin Analogs, Cabergoline, Metformin and Tramadol. Her laboratory investigations with treatment were controlled at that time. After that, the patient stopped taking her medications a couple of years ago by her own decision, because of her desire to get pregnant as she has been told that the medications are teratogenic. As a result of that, her laboratory investigations got worse. Two Plastic surgeries were performed for her left face deformity in 2014 and in2015.
On physical examination, she had scattered brown pigmentation on her face and body and there was left asymmetrical face mass with asymmetrical eyes shape (Figure 1). Her Body Mass Index was 29.9 (Overweight).

The patient was admitted that same day for further investigation. Laboratory investigations on admission (complete blood count, kidney function test, liver function test and urine analysis) were normal except for abundant white blood tests in the urine analysis. Thyroid Function Tests, Growth Hormone and Insulin-like Growth Factor 1 were ordered but were not available. Pelvic magnetic resonance imaging was performed and showed severely deformed vertebral bones as well as the sacrum, associated with scoliosis. An orthopedic consultant advised cesarean section, due to her affected vertebral bones that will be at risk of fracture if she will deliver vaginally. She was advised to do proper imaging and rehabilitation postpartum. An endocrinologist was consulted for evaluation and they recommended further follow up after delivery at the outpatient clinic.

The patient underwent an elective cesarean section at 39 completed weeks and the anesthesiologist preferred general anesthesia over spinal anesthesia due to the deformities of the vertebrae and the presence of scoliosis. The outcome was a healthy female weighing 3500 gm. Hospital course went smoothly without any complications. The patient and her newborn were discharged on the 2nd day after surgery, and postpartum follow ups were uneventful.

Figure 1: the photo of the patient's face (after taking permission from her to publish it) 1/6/2019

DISCUSSION

Women with polyostotic fibrous dysplasia have high prevalence of abnormal uterine bleeding which may require blood transfusion and therapeutic hysterectomy at unusually younger age (4). First line management for such cases who do not desire pregnancy is levonorgestrel intrauterine device (7). There is no clear explanation in the literature of the cause of this condition in this disease. In our case, the patient had abnormal vaginal bleeding in the form of amenorrhea. The cause of amenorrhea was unknown by the patient but she had been treated with oral combined contraceptive drugs that regulated her menstrual cycle.

Effects of polyostotic fibrous dysplasia on fertility and childbearing have not been determined (5). According to a large study of cases (4), fertility is impaired in fibrous dysplasia; however the possibility remains for spontaneous conception. In our case, the patient had spontaneous pregnancy as soon as she had stopped her combined oral contraception drugs.

Effects of pregnancy on skeletal outcomes have not been decided yet. Only limited conflicting data is available of case reports of polyototic fibrous dysplasia skeletal complications during pregnancy, including increased bone pain in most cases, bone cysts aneurysms, and malignant transformation of fibrous dysplasia lesions. This has led some to surmise that pregnancy increases fibrous dysplasia activity, stating them for poor skeletal outcomes (4, 5). One of the studies has mentioned a woman who was not known to suffer from fibrous dysplasia until she became pregnant and experienced a dramatic reactivation of the symptoms (8). Another study suggested that women with disfiguring lesions have more risk of exacerbation during pregnancy (9). Pregnancy is marked by high bone turnover state. IGF-I levels may be an important determinant of bone turnover during pregnancy. Elevated bone turnover may explain bone loss during pregnancy (10, 11). Interestingly in our isolated fibrous dysplasia case, despite stopping her medications, the generalized bone pain had decreased after she got pregnant. There was no increase in her face disfiguring lesion. There was no increase in serum glucose levels.

The mode of delivery was mentioned in a case report that was published in 1958. It included two cases and both patients had the classic abnormalities of polyostotic fibrous dysplasia which were unilateral cystic changes within the skeleton, and associated unilateral pigmentation in the skin. They were delivered vaginally without any damage to the architecture of the pelvis (6). In our case, a cesarean section was done as recommended by the orthopedic consultant after evaluating her case.

CONCLUSION

Polyostotic fibrous dysplasia/McCune Albright syndrome is rarely encountered in pregnancy. In our case report, it did not have any adverse effect on the pregnancy and pregnancy did not worsen the

condition. However, we recommend pelvic and spinal imaging to determine the mode of delivery as well as the type of anesthesia if cesarean section is needed.

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