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PREGNANCY IN PROGRESSIVE OSSIFYING FIBRODYSPLASIA: CASE REPORT

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BACKGROUND

Progressive ossifying fibrodysplasia (POF) is a rare autosomal dominant disorder that results from spontaneous mutation of bone morphogenic protein1 receptor ACVR1. This pathology is characterized by congenital hallux malformations and ectopic ossification of the connective tissue in the skeletal muscles and ligaments. It may lead to progressive immobility and cardiorespiratory complications, with high morbidity and mortality. Trauma stimulates flares. Therefore, invasive procedures with diagnostic purpose should be avoided. Since smooth muscle is not involved in POF, pregnancy is possible, although it represents a risk to maternal-fetal life. However, pregnancy is a rare occurrence in these cases, with few reports of advanced gestation.

CASE REPORT

Female, 41 years old, begins outpatient follow-up at the medical clinic, presenting pain and increased volume of hands and feet. (Figure 1). Born with right breast agenesis and toenail malformation, she developed, at 4 years old, signs of increased lower limb volume. Diagnosis of lymphedema was suggested. Gynecological history: menarche (11 years old), menstrual regularity, 3 pregnancies (3 cesarean), without abortion. Surgical menopause at 38 years (myomatosis uterine). At 21 years old, after her first gestation, joint manifestations got worse (low back pain, polyarthralgia, difficulty in ambulation and limitation of movement arches). In 2012, the patient was referred to rheumatologic evaluation. She presented episodes of increased articular volume and soft tissues, accentuated in her hands, feet and right hip, with phlogose, culminating in heterotopic calcifications. These calcifications were confirmed by radiological findings, typically of POF. (Figure 2) Corticosteroid intravenous cycles was prescribed for treating flares, decreasing phlogistic process and partially delaying soft tissue calcification. Afterwards, oral bisphosphonate was introduced empirically.

CONCLUSION

There are few reports in literature that describe pregnancy in patients with this pathology. It is known that POF is a progressive and limiting condition to life. Gestation, even if possible, may accelerate disease progression. In the presented case, the patient had successful in 3 pregnancies. Her children were born healthy by cesarean delivery, although there is a 50% chance of genetic inheritance for this condition.

Clinic manifestations in FOP are indispensable, for assertive and early diagnosis of POF, since invasive measures should be avoided. After diagnosis, patients should access genetic counseling and be encouraged to undertake conscious family planning.