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CASE REPORT

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Telangiectatic osteosarcoma initially presenting as right popliteal vein thrombosis in pregnancy: A case report

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Abstract

Background: Telangiectatic Osteosarcoma (TO) is a rare variant of osteosarcoma. The rarity of TO and its similar initial presentation with Deep Vein Thrombosis (DVT) often leads to missed diagnosis or misdiagnosis during the primary evaluation.

Case presentation: A 30-year-old Para 1+0 Gravida 2 presented with right lower limb swelling with knee effusion, right lower leg thrombosis, and TO at 30 weeks gestation to the Kenyatta National Hospital (KNH). Serial limb measurements, analgesics for lower limb pain, fetal heart rate, and movement monitoring were done. Disarticulation at the hip joint was scheduled, but the case declined further care, opting for expectant management. She had an intrauterine fetal demise and delivered a macerated stillbirth at 31 weeks of gestation. She was admitted for one day in the labor ward, discharged, and was lost to follow-up.

Conclusion: Telangiectatic osteosarcoma can present as right popliteal vein thrombosis.

Keywords: Telangiectactic Osteosarcoma, Venous Thromboembolism (VTE), Spontaneous pregnancy loss, Osteosarcoma

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Introduction

Osteosarcoma (OS) is a rare cancer of long bones, predominantly the distal femur, proximal tibia, and proximal humerus. It presents with a dull or aching pain with or without a pathologic fracture, irritation of periosteum, limping, impaired ambulation, and penetration of tumor in the cortical bone, commonly in the second decade of life (1). The annual incidence of osteosarcoma is 5.6 in 1 000 000 in adolescents (<15 years), 3.4 in 1 000 000 in individuals aged ≤24. Osteosarcoma is rare in under-fives, while individuals between 25 – 59 years bear the least risk at 1.9 in 1 000 000 (2). The survival rate is 60-70% if non-metastatic, but systemic spread carries a greater risk of mortality at 70% (3). Telangiectatic Osteosarcoma (TO) is a rare variant of OS accounting for 2-12% of OS cases. On x-ray evaluation, TO appears as a lytic mass lesion with osteoid and cystic spaces with blood and thin septa, often in the metaphyseal section of a long bone. Serum Alkaline Phosphatase (AKP) may be mildly elevated or normal. Matrix mineralization, new bone formation, cortical destruction, and bone lysis are common radiographic findings (4). Clinical outcomes are good when Lactate Dehydrogenase (LDH) levels are normal and the tumor is small. However, it can be misdiagnosed as Aneurysmal Bone Cysts (ABC) and refractive Deep Vein Thrombosis (DVT) or missed during clinical evaluation, leading to adverse obstetric outcomes (4-5).

This is a case of a 30-year-old pregnant woman diagnosed with possible Venous Thromboembolism (VTE) and later TO of the right lower limb, with subsequent spontaneous pregnancy loss.

Case presentation

A 30-year-old Para 1+0 Gravida 2 at 30 weeks + 6 days gestation presented to the Kenyatta National Hospital (KNH) with TO. She was initially diagnosed with VTE of the right lower limb. On clinical examination, her right lower limb was warm and swollen. The calf circumference was 37 cm versus 30 cm in the asymptomatic limb. The calf circumference was measured 10 cm below the tibial tuberosity. She had a limping gait and impaired ambulation. She gave no history of known food or drug allergies, blood transfusion, Human Immunodeficiency Virus (HIV), diabetes mellitus, hypertension, varicose veins, or a known family history of VTE. She was a non-smoker, a non-alcoholic but sat for eight hours per day at work, relieved by walking. The fetus was in cephalic presentation with a fundal height corresponding to 30 weeks of gestation and had good fetal tone, normal amniotic fluid index, and normal regular heart rate of 140 Beats Per Minute (BPM) on ultrasonography. Right lower limb venous Doppler ultrasound revealed an effusion. Knee joint fluid aspiration and a peri-joint biopsy revealed fragmented tissues of skin fibrous tissue, bone, osteoid, skeletal muscle, and markedly hypertrophic cells with no hypertrophic pattern, but the diagnosis was inconclusive. A greyscale color Doppler evaluation of the common femoral, superficial femoral, popliteal, and deep calf veins revealed an echogenic thrombus occluding the right's whole diameter popliteal vein. Right knee joint effusion and edema of the subcutaneous tissue were also seen and diagnosed as knee joint effusion and right lower leg VTE. The symptomatic limb was elevated. She was managed with analgesics, antibiotics, and 60 mg of enoxaparin daily for seven days.

The patient was scheduled for multiplanar multi sequential Magnetic Resonance Imaging (MRI) and intralesional open biopsy evaluation. Coagulation tests were as follows: Prothrombin Time (PT) (18 seconds), activated Partial Thromboplastin Time (aPTT) (31 seconds), International Normalized Ratio (INR) (1.29), and Clotting Time (CT) (30 seconds). Her Hemoglobin (Hb) was 13.3 g/dL, White Blood Cells (WBC) 6.96 X 10^9/L, Platelets 307 x 10^9/L, and neutrophils 4.72 X 10^9/L. The Alkaline Phosphatase (ALP) was markedly elevated at 1038 U/L (Normal 37-116U/L). Urinalysis findings were: severe levels of ketones and leukocytes, moderate proteins, and the urinary Specific Gravity (SG) was 1.025. A diagnosis of asymptomatic bacteriuria was inferred. Magnetic resonance imaging showed a large corticoid-based tumor of low signal on all sequences with widespread infiltration of marrow and adjacent soft tissue with heterogeneous enhancement after intravenous contrast. Subcutaneous tissue swelling, joint articulation congruence, and tibial condyles were seen, and a distal femoral malignant tumor was secluded for surgical excision. After an intralesional open biopsy, the specimen was sent for histopathological analysis, which reported a tumor with sheets of round, oval, and spindling epithelial cells with a distinct and scanty amphophilic cytoplasm with mild pleomorphism, collagenous hyalinization, and trabeculae of osteoid with a lace-like pattern, suggestive of osteosarcoma, the telangiectatic type. The patient was scheduled for disarticulation at the hip joint, following enhanced counseling. However, she declined further care, and opted for expectant management awaiting delivery. The management involved daily serial limb measurements, analgesics administration for lower limb pain, fetal heart rate, and movement monitoring. The case had a macerated stillbirth at 31 weeks of gestation, approximately seven days after admission, delivered vaginally after labor induction. The patient was admitted for one day at the labor ward, discharged home, and was lost to follow-up.

Discussion

This case's findings confirm that VTE may present with concurrent TO. Because the symptoms of VTE and TO are similar. The risk of TO is higher among the youth who are immobile or sit for long hours at work. Painful lower leg swelling, deranged PT and CT, markedly elevated ALP, and high levels of ketones, leukocytes, and protein in urine are the significant risk factors for TO. Musculoskeletal cancers such as telangiectatic osteosarcoma induce hemostatic complications such as VTE by creating a procoagulant state. Expression of coagulation factors such as Plasminogen Activator Receptor (uPAR) and Tissue Factor (TF) is higher with sarcomas, which can directly invade and compress blood vessels as the tumor grows, causing thrombosis (6). Therefore, whenever clinical evaluation reveals possible VTE diagnosis, screening for musculoskeletal sarcomas such as telangiectatic osteosarcoma should be prioritized.

The etiology of distal femoral telangiectatic osteosarcoma is unknown. However, clinical signs mirror those of osteosarcoma, including lower leg pain, which causes a limping gait or impairment of movement, subcutaneous swelling on clinical examination, and deranged coagulation parameters, especially clotting time and

prothrombin time, elevated ketones, and leukocytes in urine. The presence of a large corticoid tumor on the distal femur with widespread infiltration, spindling epithelial cells, or collagenous hyalinization in MRI and histopathology analysis were also noted, which mirror findings reported elsewhere (7-8). Phatak et al. (2005) reported pain and swelling in a 21-year-old boy with telangiectatic osteosarcoma (7). Another case reported pain, swelling, and a soft cystic mass with osteoid-producing stromal cells in a missed diagnosis of a 15-year-old with TO of the radius (8). A higher index of suspicion for TO should be indicated when a patient presents with such symptoms and diagnoses are inconclusive.

If hematologic disorders such as DVT or VTE are present, surgical resection of non-metastatic OS tumors and thrombosis removal is recommended. However, when patients present with metastatic sarcomas and tumors that do not qualify for surgical resection, disarticulation, and removal of the affected limb(s) should be prioritized, even though the survival rate with metastasis is low at 20% (9). Furthermore, the invasive nature of disarticulation, resistance, and neglect should be anticipated, as was the case. Neglecting treatment might have led to a macerated stillbirth. Even though a causal relationship between sarcoma and perinatal mortality was not established medically or statistically, osteosarcomas symptoms have been shown to worsen during pregnancy, hinting at a positive association (10).

Conclusion

Telangiectatic osteosarcoma can present as right popliteal vein thrombosis.

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