Severe Knee Deformity in a Cyanotic Patient: A Case Report

Zahra Khajali MD¹, Arash Hashemi MD², Ashkan Hashemi, MD³

Abstract

Hypertrophic osteoarthropathy is one of the rheumatologic complications in cyanotic heart disease. The severity of involvement depends on the degree and duration of a right-to-left shunt. The mechanism is related to the bypass of mediators from the lung and entrapment in the limbs. These mediators induce local cell proliferation in the bone and soft tissue. Herein, we report a cyanotic patient with severe involvement and deformity in his knee (Iranian Heart Journal 2012; 13 (1):46-49).

Keywords: hypertrophic osteoarthropathy■ Truncus arteriosus ■ Eisenmenger complex

Hematologic complications related to cyanotic heart disease are mostly gout or especially hypertrophic osteoarthropathy, which is the main cause of arthralgias, and bone pain in patients. Its mechanism is probably in relation to megakaryocytes released from the bone marrow. In a patient with a right-to-left shunt, these megakaryocytes bypass the lung and then induce local cell proliferation.¹ We report the case of a 21-year-old male, referred to our center with cyanosis, history of TA, and a unique knee deformity. The patient underwent extensive evaluation in our center and ultimately was diagnosed with hypertrophic osteoarthropathy and the Eisenmenger syndrome. In this article, we highlight joint and bone anomalies in reaction to the Eisenmenger and shunt physiology.

Case report
The patient was a 21-year-old man, who was referred to our center with a diagnosis of truncus arteriosus and pulmonary hypertension commencing 15 years previously. In our evaluations, the patient was cyanotic and had no syndromic features.

Fig. 1. Cyanosis and clubbing in the upper and lower extremities and bilateral knee joint deformity
In physical examination, positive findings were clubbing and cyanosis in the fingers in the upper and lower extremities. In cardiac examination, we found a single S2, best heard in the right upper intercostal space; 3/6 systolic and moderate duration decrescendo diastolic murmur; bounding pulse at peripheral; bilateral symmetric knee deformity without restricted motion of the knees; tenderness; and swelling or erythema on the knees (Fig. 1).

Best possible cause of this deformity was osteoarthropathic changes due to a prolonged right-to-left or bidirectional shunt and/or the Eisenmenger physiology. Ultimately to rule out other differential diagnosis, orthopedic and rheumatologic consultations were done.

Electrocardiography showed normal sinus rhythm with biventricular hypertrophy and tall R wave in V1 and incomplete RBBB pattern.

In echocardiographic examination, we encountered situs solitus, mild LV dilation and mild LV dysfunction, and mild RV dysfunction and RV enlargement. Common AV valve was quadricuspid, and there was absence of PA origination in the ventricles. Main PA was branched from the postero-lateral aspect of a common trunk and then divided to RPA and LPA. The patient had a left-sided aortic arch, and there was severe regurgitation via the common AV valve and mild to moderate MR and moderate to severe TR.

Echocardiographic findings were consistent with persistent truncus arteriosus anomaly (type A1 Van Praagh).

The patient was not a good surgical candidate at this point, and he was discharged with recommendation of close medical follow-up.

**Discussion**

Hypertrophic osteoarthropathy is a syndrome with characteristic finding of extremities enlargement due to bone and soft tissue proliferation. It is usually associated with painful joints. The major etiology of this syndrome is secondary to pulmonary...
problems, although other conditions such as cyanotic heart disease cause this syndrome.\textsuperscript{2,3} and \textsuperscript{4} The clubbing of the digits is almost always seen as a constant finding in hypertrophic osteoarthropathy and is associated with cyanotic heart disease.\textsuperscript{5} The concept of hypertrophic osteoarthropathy formation in cyanotic heart disease is the breakaway of the systemic mediators from inactivation in lungs. One of the most important mediators in this abnormality is platelet-derived growth factor (PDGF), which induces new bone and tissue formation.\textsuperscript{6,7} This syndrome is rarely seen in cyanotic patients, and the severity of involvement depends on the degree and duration of the right-to-left shunt.\textsuperscript{1} In our patient, the intensity of involvement was drastic, and the patient suffered from severe chronic knee pain. X-ray is helpful to the diagnosis of this syndrome. X-ray typically shows periosteal reaction around the distal part of long bones, and another finding is the calcification of the soft tissue around the involved bone. The involvement is usually symmetric and more frequent in the radius and fibula and to a lesser degree in the femur. Given the condition of our patient, analgesics were prescribed to control pain and the patient was scheduled for heart and lung transplantation in the future.

References


5. GEORGE E. MCLAUGHLIN.; DANIEL J. MCCARTY, JR.; and D. F. DOWNING. Hypertrophic Osteoarthropathy Associated with Cyanotic Congenital Heart DiseaseA Report of Two Cases.Annals of Internal MedicineSeptember 1, 1967 vol. 67 no. 3 Part 1 579-587.
