CASE REPORTS

Polycythemia Rubra Vera Presenting as Unilateral Clubbing Due to Left Subclavian Artery Thrombosis

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Abstract

We report the case of a 29 year old male who presented with burning pain in the tips of fingers of left hand and recurrent episodes of amarausis fugax.Examination revealed polycythemia, unilateral clubbing with positive Adson's test in left upper limb. Evaluation revealed thrombosis of the left subclavian artery in CT angiography. Polycythemia rubra vera was found to be the underlying cause.

Introduction

Clubbing is an important clinical sign in medicine and often unmasks the presence of a hidden disease. Clubbing is generally bilateral and occurs unilaterally in rare conditions. Unilateral clubbing is usually associated with local vascular lesions of the arm, axilla, and thoracic outlet and with hemiplegia. Though myeloproliferative neoplasms like polycythemia vera can cause multiple thrombotic manifestations, presentation as unilateral clubbing is uncommon.

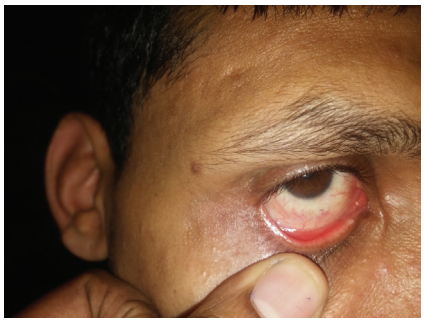

Fig. 1: Congested palpebral conjunctiva due to polycythemia

Case Report

A 29 year old male from Kozhikode, shopkeeper by occupation with no comorbidities or addictions presented with history of burning pain in fingers of left hand for 2 months duration.It was associated with difficulty in doing fine activities with left hand such as gripping an object, buttoning shirts. There was no history suggestive of proximal muscle weakness in the same limb. He also gave history of recurrent episodes of transient loss of vision in the left eye which recovered in 20-30 minutes. There was no history of headache, altered behaviour, seizures, other cranial nerve dysfunction. No history of oral ulcers, Raynaud's phenomenon, photosensitivity, hair loss. No h/o chest pain, breathlessness, palpitations, syncope. Examination revealed conjunctival congestion (Figure 1) with facial flushing and unilateral clubbing of left hand (Figures 2 and 3). Pulse was 76/min, regular rhythm, normal volume, condition of vessel wall. There was no radiofemoral delay or asymmetry of pulses between the upper limbs. Blood pressure in both upper limbs were 120/80 mm Hg. Adson's test was positive in left upper limb. Optic fundi revealed evidence of branch retinal artery occlusion in left eye (Figure 4). Rest of the neurological

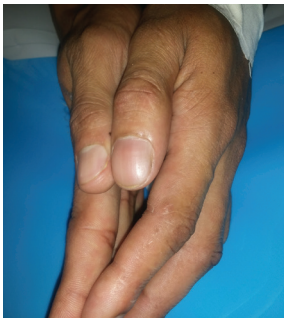

Fig. 2: Clubbing of left thumb

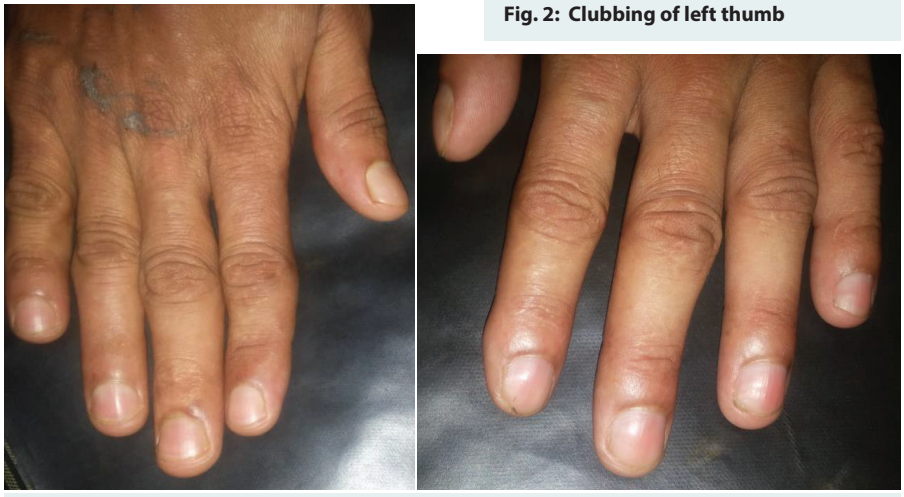

Fig. 3: Left hand showing clubbing. Right hand has no clubbing

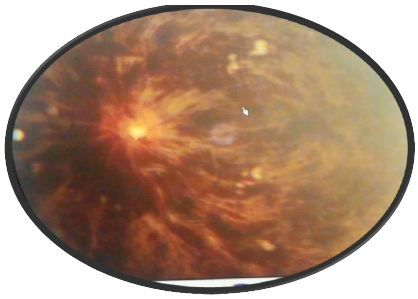

Fig. 4: Optic fundi showing branch retinal artery occlusion

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Received: 15.05.2016; Revised: 24.10.2016; Accepted: 21.12.2017

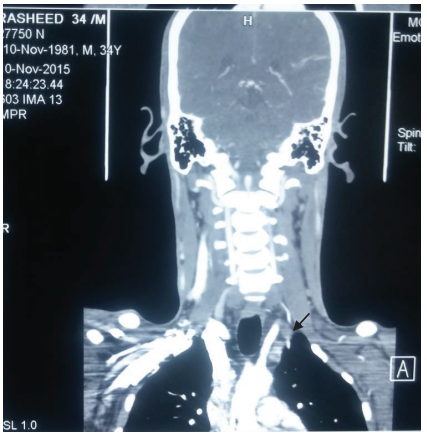

Fig. 5: CT angiography showing thrombosis of left subclavian artery (arrow)

examination was within normal limits. Cardiovascular, respiratory and gastrointestinal examination were also within normal limits.

Investigations showed Hb-21 g/dl. TC-17000/cmm, Platelet count-9.33 L/cmm. Hct-54 % ESR-5 mm in 1st hr,RBS-96 mg/dl, Blood urea -18 mg/dl, Serum creatinine-0.9 mg/dl, Total protein-7 g/dl, albumin-4 g/dl, SGOT/SGPT-13/43 IU/L. Urine routine examination was within normal limits. Electrocardiogram showed normal sinus rhythm with no abnormal changes. Chest x-ray was within normal limits and there was no evidence of cervical rib. Computed tomography was done to exclude any cerebro vascular pathology, was within normal limits. JAK2 gene mutation study by real time PCR was detected and serum erythropoietin was low-3.64 milliIU/ml (4.30-29) thus confirming the diagnosis of polycythemia rubra vera. CT angiography of left upper limb vessels revealed thrombosis of left subclavian artery (Figure 5). He was treated with anticoagulation, and venesection was given. He improved symptomatically and is kept under follow up.

Discussion

Thrombotic manifestations in myeloproliferative neoplasms are common but presentations as unilateral clubbing due to a local vascular cause is quite rare. Although the mechanisms involved in this hypercoagulable state are unclear, abnormalities in blood viscosity, platelets, and leukocytes have been implicated.[1] Causes of unilateral clubbing include local vascular lesions of the arm, axilla, and thoracic outlet and with hemiplegia. Vascular aneurysm, of the subclavian artery, innominate artery, syphilitic aortitis, carcinoma of upper lobe of lung are other reported cases of unilateral clubbing.[2] Other causes of Subclavian artery thrombosis include arterial thoracic outlet syndrome caused by a cervical or anomalous first rib.[6] Compression of the subclavian artery by these bony abnormalities produces stenosis, post-stenotic dilatation, formation of aneurysms and mural thrombosis. It can occur as a consequence of trauma and can be exacerbated by repetitive movements such as those performed by athletes. Thrombus can form over atheromatous plaques located within the subclavian artery. Large vessel vasculitis can involve the subclavian artery causing wall thickening, fibrosis, stenosis and thrombus formation.

Treatment of polycythemia vera should include phlebotomy to keep haematocrit below 45 percent and low dose aspirin. Anticoagulation should be initiated in the setting of a thrombotic event though there are no randomised controlled trials regarding duration of treatment.

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