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CASE REPORT

A case of multivessel PTCA in achondroplasia patient

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1. A case of multivessel PTCA in achondroplasia patient

Dwarfism may be due to several musculoskeletal and hormonal growth disorders. The most common cause is considered to be achondroplasia, a condition due to a mutation affecting the fibroblast growth factor receptor (FGFR) gene 3. Achondroplasia occurs with equal frequency in males and females. It is inherited in an autosomal dominant manner.[1] At least 80% of cases result from a random new mutation. In sporadic cases, a paternal age older than 36 years is common.[2]

Despite an estimated prevalence is 1:25,000 in the general population, [3] there is little literature concerning the diagnostic and treatment challenges faced by doctors dealing with CAD in such patients requiring myocardial revascularization.

2. Case report

A 46-year-old Iraqi male with achondroplasia presented with intermittent rest angina and dyspnoea for last 1–2 months relieved by sublingual nitrate. His height was 85 cm, his weight was 70 kg, and he had severely atrophic limbs with kyphoscoliosis (Fig. 1). He was having hypertension and dyslipidemia as risk factors. The electrocardiogram showed Q wave in inferior leads with T wave inversion. Echocardiogram revealed LV ejection fraction of 45–50% with RWMA in inferior territory.

CAG was planned via radial artery route in view of femoral artery access issues and to avoid local bleeding complication. Left radial artery access was taken as preferred route in view of anticipated tortuosity. There was severe radio-brachial tortuosity (Fig. 2) and great difficulty was encountered in tracking diagnostic catheter. Angiography revealed TVD with critical disease in LAD, major diagonal, major OM and RCA proximal, which was 100% occluded (Figs. 4, 7 and 9). Surgical revascularization was the initial plan. Heart team was involved. CABG required multiple grafts but in view of severe musculoskeletal deformity, surgeons were not optimistic of suitable grafts. So PTCA was planned after taking informed consent. Patient was preloaded with Ticagrelor 180 mg, aspirin 325 mg and atorvastatin 80 mg. Unfractionated heparin was used as the anticoagulant. PTCA +S with DES were done to RCA, LCX-OM2 and proximal to mid LAD, and POBA to proximal PLB and diagonal arteries.

PTCA was done via right femoral artery access using 7 Fr femoral sheath .Ultrasound guided femoral artery puncture was done. The RCA was cannulated with 7 Fr, 3.5 curve Judkins right guiding catheter with the catheter tracked over an amplatz super stiff wire because of the severe aortic-iliac tortuosity (Fig. 3). Lesion was crossed with 0.014″ whisper guidewire with the support of a 1.25 × 15 mm balloon. A 3.5 × 36 mm DES was deployed across the lesion in RCA at 14 atm after pre-dilatation. After dilating proximal 100% RCA lesion, another critical lesion was evident in proximal segment of a very large PLB. Prolonged low pressure dilation to proximal PLB lesion was done with 2.5 × 10 semi-compliant balloon at 6 atm. Distal TIMI 3 flow was achieved at the end of the procedure (Figs. 5 and 6).

Then LCA was cannulated with 7 Fr, 3.5 curve EBU guiding catheter. A 0.014″ whisper guidewire was then used to cross the major OM lesion. A 2.5 × 14 mm DES was deployed

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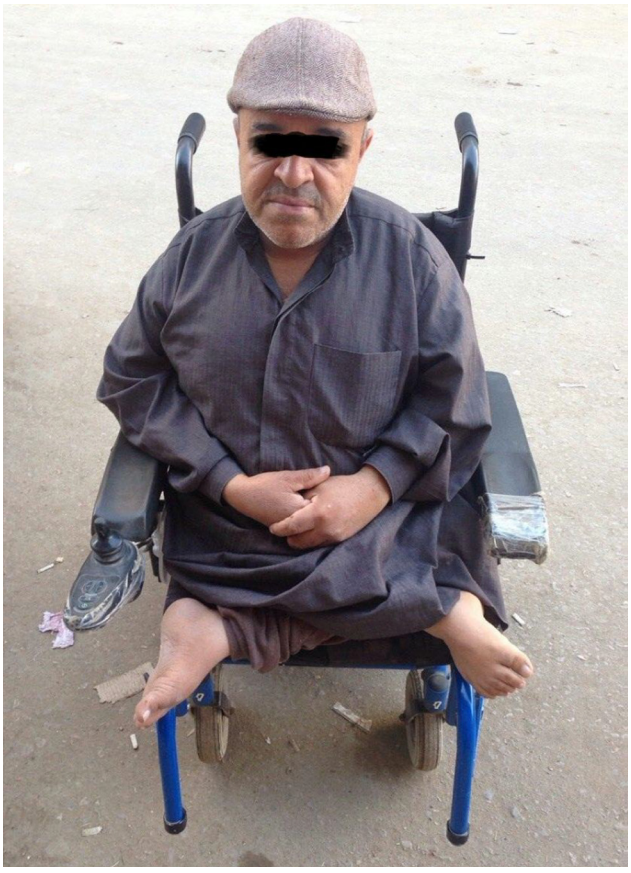

Figure 1 The patient of achondroplasia.

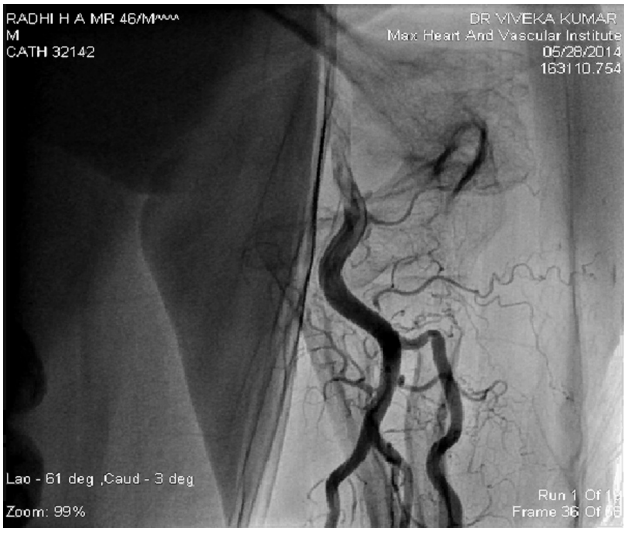

Figure 2 Radio-brachial artery tortuosity.

across the lesion from LCX-OM at 14 atm after pre-dilatation. Post-dilatation distal TIMI 3 flow was achieved (Fig. 10).

Then whisper wire was crossed across diagonal lesion and 0.014″ BMW wire was crossed across LAD lesion. Balloon dilatation was done to proximal diagonal lesion with 2.5 × 10 semi-compliant balloon at 8 atm. A 2.5 × 33 mm

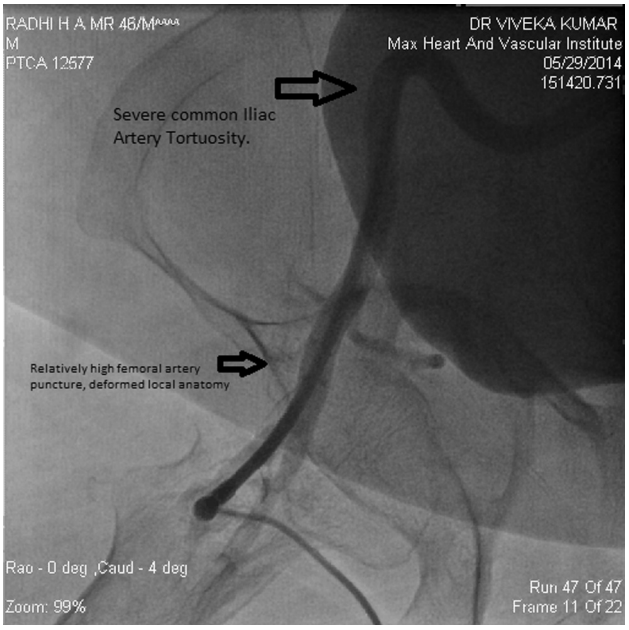

Figure 3 Common Iliac artery tortuosity.

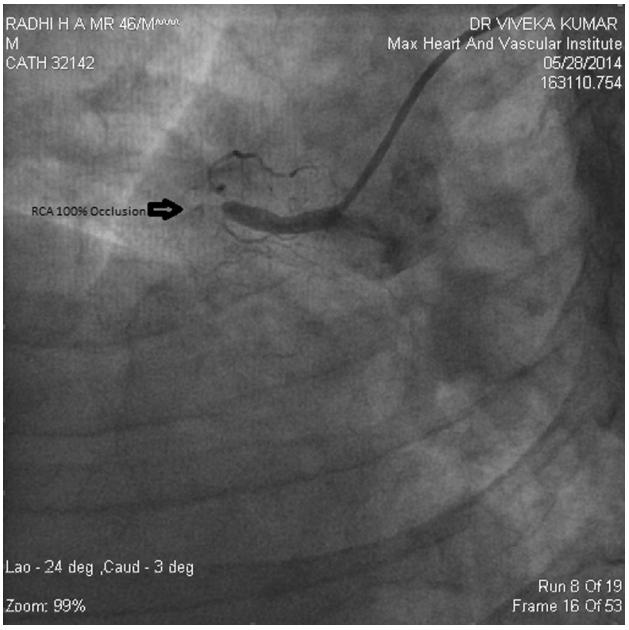

Figure 4 RCA proximal 100% occlusion.

DES was deployed across the proximal-mid LAD lesion at 14 atm after pre-dilatation across major diagonal. Post dilatation distal a TIMI 3 flow was achieved (Fig. 8). Right femoral artery puncture was closed with **Perclose Proglide**® (Abbott Vascular) suture mediated closure device after completion of the procedure. Later course in the hospital was uneventful and he was discharged in a stable condition on day 3 of hospitalization. After discharge, the patient was placed on aspirin, ticagrelor, statin, beta-blocker and ACE inhibitor (ramipril).

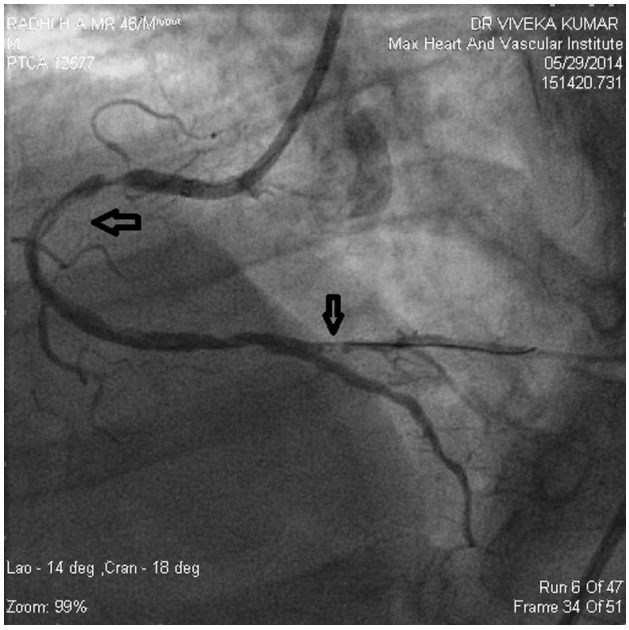

Figure 5 After Predilatation to RCA.

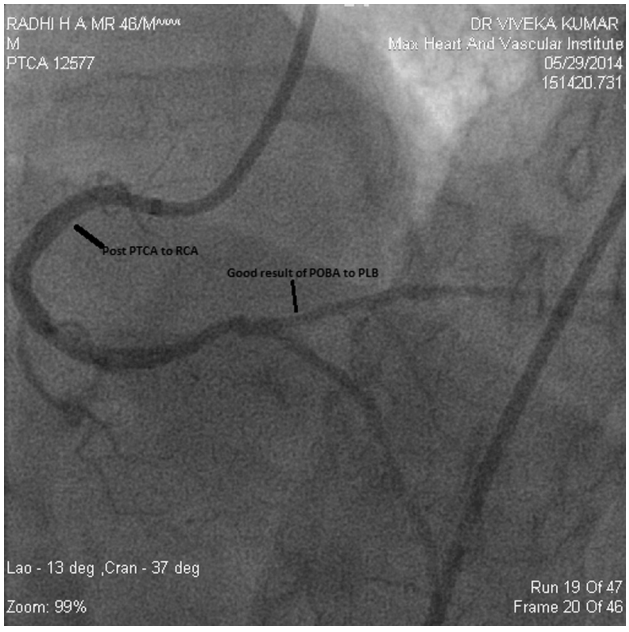

Figure 6 RCA – final result.

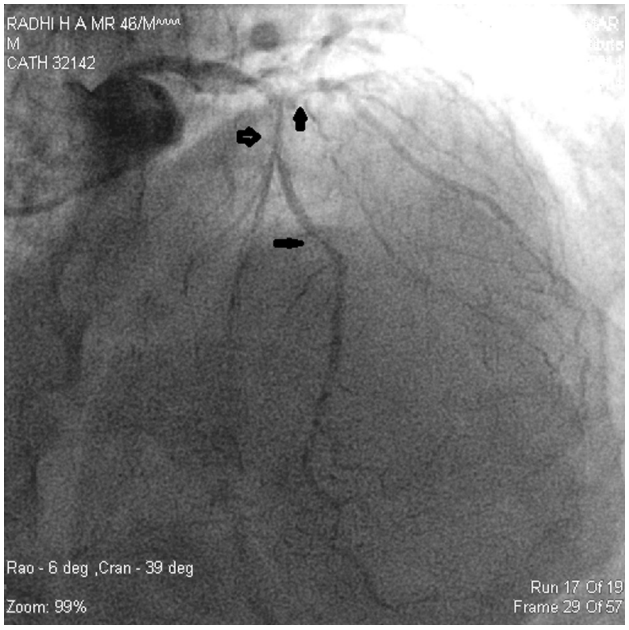

Figure 7 Critical disease in LAD and diagonal.

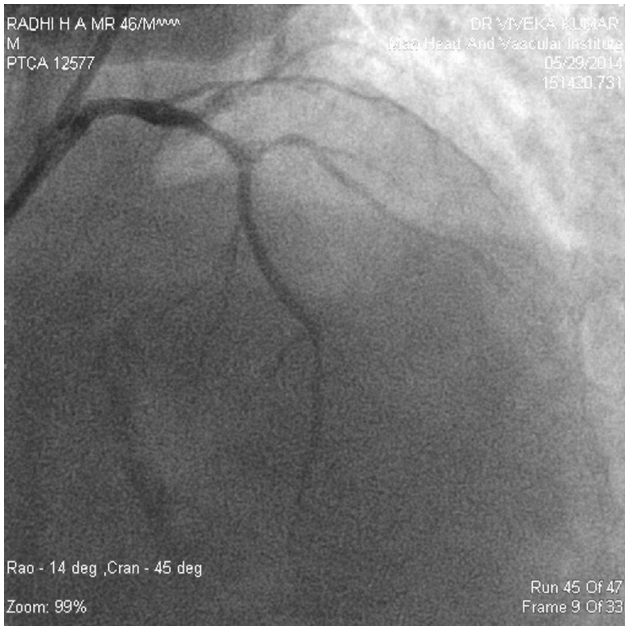

Figure 8 LAD – Diagonal: Final result.

3. Discussion

Mortality in general in these individuals for all age groups is 2.27 times more than that of the general population, cardiovascular problems being the most frequent causes of death in persons aged 25–54 years[3]. Apart from the traditional risk factors, there seems to be other occult genetic or other unknown factors responsible for this increased risk. No large cohort studies have been done to delineate these additional factors till date. There is dearth of literature regarding the technical aspects of coronary intervention in this group of individuals. There are a few reports of coronary artery bypass surgery in these patients,[4,5] and a single report on PCI to RCA as rescue PCI post thrombolysis[6]. Option of CABG in the setting of multivessel CAD in an achondroplastic patient may be ruled out because of non-availability of adequate grafts as in our case. However there is a report of successful CABG after a phlebography of the limbs revealed adequate saphenous veins[5], but the patient had a height of 137 cm height, in our case patient's height was 85 cm only. So in such short patients with multivessel CAD multivessel PTCA may be the only option to achieve myocardial revascularization. To our knowledge, this is the

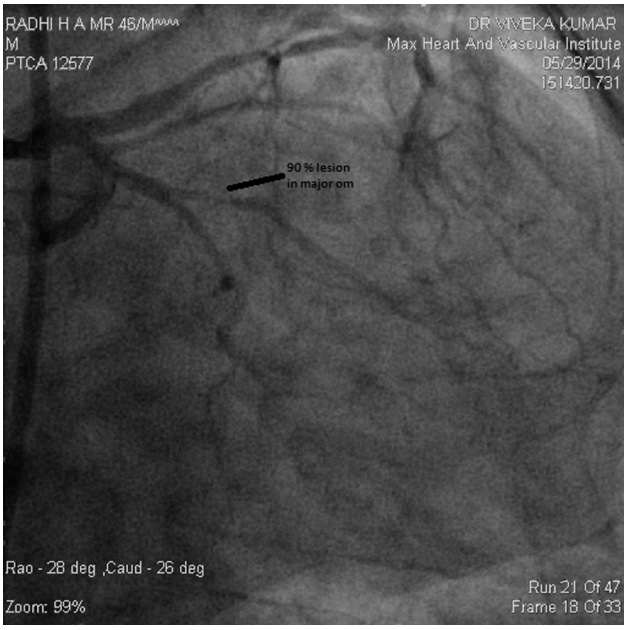

Figure 9 Critical lesion in major OM.

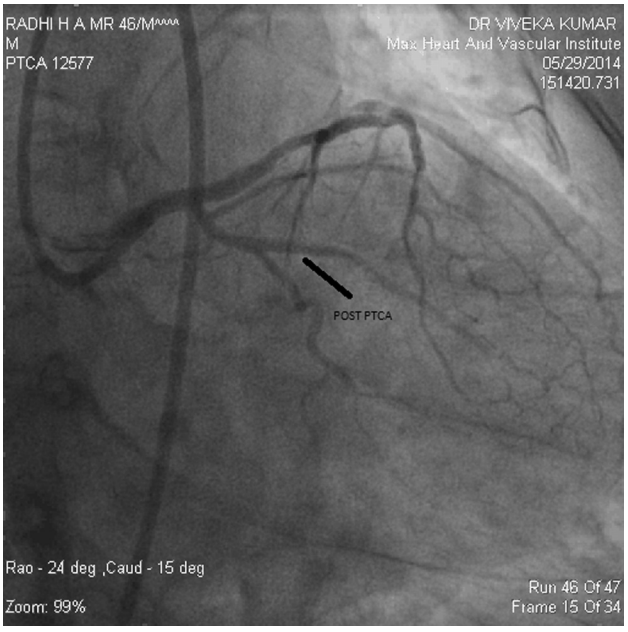

Figure 10 LCX-OM: Final result.

first case report of mutivessel PTCA in an achondroplastic patient. Radial intervention may prove to be particularly challenging in such cases, but with proper hardware and adequate skills it may still be feasible to do noncomplex PTCA. Local bleeding complication was minimized by using USG guided femoral artery puncture and post procedure use of femoral artery closure device. Our case demonstrates that patients with achondroplasia can safely undergo multivessel coronary angioplasty without additional risk.

4. Conclusion

Achondroplastic patient having multivessel CAD poses a great challenge for myocardial revascularization. CABG as an option may be limited by non-availability of normal saphenous vein grafts in such patients having severely dysmorphic limbs. Multivessel PTCA can be safely performed in patients with dwarfism due to achondroplasia with the available technical knowhow and hardware. Further studies need to be done to look into the increased risk of CAD in such population.

Conflict of interest

There is no conflict of interest.

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