CASE REPORT

Klippel-Trenaunay Syndrome and Radial Artery Coronary Graft Spasm

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ABSTRACT

A 75-year-old woman with known diagnosis of Klippel-Trenaunay syndrome presented with acute onset of chest pain, dyspnea and elevated cardiac enzymes. She had triple vessel coronary artery disease on subsequent coronary angiography. Given the unavailability of venous conduits secondary to lower extremity varicosities, coronary artery bypass grafting with radial and internal mammary arterial grafts was carried out. The radial artery graft went into spasm two days later and required intracoronary vasodilators to relieve the spasm. The patient remained hypotensive and finally expired.

Key words: *Klippel-Trenaunay syndrome (KTS). Coronary artery bypass graft surgery (CABG). Percutaneous intervention (PCI). Quantitative coronary arteriography (QCA). Radial artery graft.*

INTRODUCTION

Klippel-Trenaunay syndrome is also known as congenital dysplastic angiectasia, angio-osteo hypertrophy syndrome and Klippel-Trenaunay-Weber syndrome. This rare congenital disorder, first described in 1900, is characterized by port wine stains, hypertrophy of bone and soft tissues and extremity vascular malformation.[1-5] Vascular malformation may be mixed involving capillary, arterial, venous or lymphatic systems mainly in extremities.[1-3] There are various hypothesis regarding arteriovenous malformations in Klippel-Trenaunay Syndrome (KTS). These include mesodermal defect during embryogenesis causing open microscopic arteriovenous communications with vascular and soft tissue malformation in the affected limb,[4] as well as angiogenic malformation involving an angiogenic factor VG5Q with t (5; 11) chromosomal transcription implicated in extremity vascular malformations. We present here a rare case of radial artery graft spasm in a patient with Klippel-Trenaunay syndrome in whom the internal mammary arterial graft was unaffected after coronary artery bypass graft surgery.

CASE REPORT

A 75-year-old woman with Klippel-Trenaunay Syndrome (KTS) presented to emergency centre with chest pain and dyspnea. The patient had bilateral lower extremity hypertrophy with vascular congestion, varicose veins and pedal edema. She had elevated cardiac enzyme. There were non-specific ST segment changes on anterolateral leads on admission ECG. Coronary angiography subsequently revealed 80% stenosis of proximal Left Anterior Descending (LAD) artery, total occlusion of left circumflex artery (LCX), and 85% stenosis of proximal Right Coronary Artery (RCA) using Quantitative Coronary Arteriography (QCA) measurements (Figure 1, A and B). The ejection fraction was 45%. The patient was referred for aortocoronary bypass surgery (CABG), but due to the KTS, she had no venous conduits available. The patient underwent PCI and Drug Eluting Stent (DES) placement in RCA and then CABG with radial and internal mammary arteries grafted to first Obtuse Marginal (OM) branch of LCX and LAD, respectively.

Two days post-CABG the patient developed a high degree AV block (Figure 2, C) and was taken to the

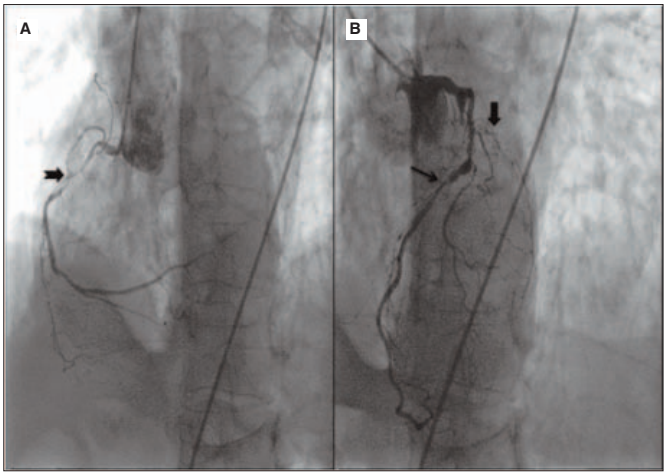

Figure 1: Pre-CABG coronary angiography (**A** and **B**) showing proximal RCA 85% stenosis (notched arrow, **A**) and 80% LAD stenosis (Thin arrow, **B**). LCX occlusion (Thick arrow, **B**) is also visible.

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cardiac catheterization lab. A temporary pacemaker was placed. An intra aortic balloon pump was inserted for mechanical circulatory support. On angiogram, she had a patent DES to RCA (Figure 2, A) and a patent internal mammary artery graft. The Radial Artery (RA) graft to OM had proximal spasm (Figure 2, A) which resolved after intragraft administration of verapamil, adenosine and sodium nitroprusside (Figure 2, B). The patient remained hypotensive, became asystolic even with temporary pacing, and died.

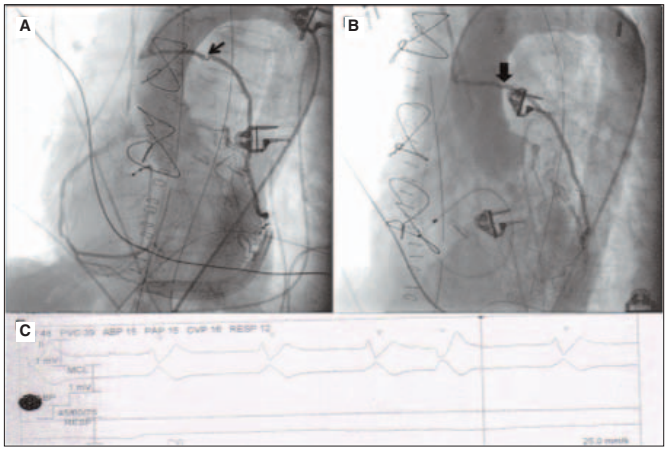

Figure 2: Post-CABG coronary angiography (**A** and **B**) showing proximal spasm of RA graft (**A**, arrow) to first OM, associated rhythm strip (**C**) and spasm resolution (bold arrow, **B**) after intracoronary vasodilator administration. Temporary pacemaker leads and DES in RCA are also visible.

DISCUSSION

Klippel-Trenaunay Syndrome (KTS) is a rare condition with involvement of skin, soft tissue and vascular system.[1-5] The vascular and soft tissue disease components in the KTS are characterized by malformation of capillary (98%), venous (72%) and lymphatic (72%) vessels along with bony and soft tissue hypertrophy (67%).[1-3] The exact etiology and pathogenesis of the KTS is unclear. Baskerville suggested that arteriovenous fistulas are due to mesodermal defect during embryogenesis causing microscopic arteriovenous communications to remain open.[4] This causes vascular and soft tissue malformation in the affected limb. Another study mentioned that intrauterine damage to the sympathetic ganglia or intermediolateral tract could cause dilated microscopic arteriovenous anastomosis leading to characteristic KTS arteriovenous malformation.[5] Xiao-li described an angiogenic malformation involving an angiogenic factor VG5Q in the KTS with increased transcription at chromosome location t (5; 11).[6]

The patient in this case report had typical features of KTS, including lower extremity varicosities with vascular obstruction and venous ulceration. The lack of venous conduits led to radial and internal mammary artery grafting. Although both Internal Mammary (IM) and Radial Artery (RA) grafts are used as arterial grafts in CABG,[7] RA graft has been shown to be more likely to have vasospasm.[6] There is also the possibility of the use of an "*in-situ*" LIMA, as opposed to a "free" radial graft to influence the occurrence of spasm. It is conceivable that the use of an *in-situ* graft can protect against spasm by preserving the vascular endothelial tone. Some studies have shown that there is reduced production of endothelium derived relaxation factors, including 6-keto-prostaglandin F1, in radial arteries as compared to the internal mammary artery.[6] Other studies reported that human RA has higher receptor mediated contractility to circulating vasoconstrictor like endothelin-1 (ET-1) and angiotensin II (A-II). ET and AII are increased in the plasma during CABG. This could lead to increased radial arterial graft spasm in the intraoperative or immediate postoperative period.

Our literature review failed to reveal any articles or case reports on arterial coronary grafts in Klippel-Trenaunay syndrome and the risk of subsequent spasm. Patients with KTS have defective angiogeneis in extremities[1-5] and it is possible that differences in extremity and truncal artery graft spasmodic events are exaggerated in KTS as compared to the general population.

A case report does not allow definite conclusions. However, it can only serve as the basis for future studies on the role of truncal versus extremity grafts in KTS and other extremity vascular malformation syndromes. Bilateral internal mammary grafts could be a consideration in these cases. There are no current randomized trials available, but observational data looks promising. This strategy could be a better option in extremity vascular malformation patients. Finally, a hybrid procedure was utilized in this case as RCA was considered the culprit lesion for hemodynamically unstable dysarrhythmias, at the time of left heart catheterization.

This case report presents a unique scenario of radial artery spasm in a Klippel-Trenaunay syndrome patient. The internal mammary graft was unaffected. Since extremity angiogenesis malformation is the predominant mechanism for vascular malformation in KTS,[1-6] we suggest radial artery grafts may be more prone to spasm than *in-situ* IMA grafts in patients with KTS and other extremity congenital vascular malformations.

REFERENCES

1. Berry SA, Peterson C, Mize W, Bloom K, Zachary C, Blasco P, *et al.* Klippel–Trenaunay syndrome. *Am J Med Genet* 1998; **79**: 319-26.
2. Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP. Klippel-Trenaunay syndrome: spectrum and management. *Mayo Clin Proc* 1998; **73**:28-36.

3. Akcali C, Inaloz S, Kirtak N, Ozkur A, Inaloz S. A case of Klippel-Trenaunay syndrome involving only upper limbs. *G Ital Dermatol Venereol* 2008; **143**:267-9.

4. Baskerville PA, Ackroyd JS, Browse NL. The etiology of the Klippel-Trenaunay syndrome. *Ann Surg* 1985; **202**:624-7.

5. Bliznak J, Staple TW. Radiology of angiodysplasias of the limb. *Radiology* 1974; **110**:35-44.

6. Tian XL, Kadaba R, You SA, Liu M, Timur AA, Yang L, *et al.* Identification of an angiogenic factor that when mutated causes susceptibility to Klippel-Trenaunay syndrome. *Nature* 2004; **427**:640-

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ERRATA

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Due to an oversight, discrepancies have occurred in the name of authoress, her e-mail address and area code in the letter to the Editor titled "Medical Education in Pakistan" published in JCPSP 2009, Vol. 19 (08): 536. The name of authoress was mentioned as Dr. Zarine S. Siddiqui instead of Dr. Zarrin S. Siddiqui, area code WA 609, instead of 6009, and missing official e-mail address.

The correct name of authoress is Zarrin S. Siddiqui, area code WA 6009 and official e-mail address zsiddiqui@meddent.uwa.edu.au, which may be corrected and read as such.

2

A discrepancy has occurred in the serial order of the name of a co-author in the article titled "Outcome of Stapled Haemorrhoidectomy versus Milligan Morgan's Haemorrhoidectomy" by Nawaid Farooque Khan *et al.* published in JCPSP 2009, Vol. 19 (09): 561-565.

The name of one of the co-authors Shahid Rasul, has been mentioned in the order as 8th author instead of 3rd. The name of third author in the above mentioned article is Dr. Shahid Rasul, which may be corrected and read as such.

We regret these mistakes.

Editor