Type A Circumferencial Aortic Dissection

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Fig. 1: (a) Left parasternal long axis view showing dissecting flaps (DF) arising from sinotubular region; (b) Apical five chamber view during diastole showing prolapse of dissecting flap (DF) in LVOT (arrow); (c) Transesophageal echocardiogram showing extension of dissection flap (DF) in dilated ascending aorta; (d) Transesophageal short axis view showing nearly circumferential nature of dissection. T- True lumen, F- False lumen.

A 40 years male presented with progressively increasing breathlessness & oedema. Transthoracic echocardiography revealed dilated aortic root and ascending aorta, two dissecting flaps (Figure 1a-DF) originating from sino-tubular region, diastolic prolapse of dissecting flap in LVOT (Figure 1b-arrow), gross aortic regurgitation, left ventricular systolic dysfunction, and severe pulmonary artery hypertension. Mitral valve, aortic arch and descending aorta were normal. Transesophageal echocardiography confirmed above findings. In addition it revealed extension of dissecting flap in the ascending aorta (Figure 1c-DF). Aortic root dimension was 50.1mm and ascending aortic dimension was 45.1mm. Short axis view confirmed circumferential nature of dissection (Figure 1d). Patient had Marfanoid habitus without ectopia lentis.

Type A aortic dissection in young persons is mostly due to genetically transmitted diseases of aortic valve or thoracic aorta eg. Marfan syndrome, bicuspid aortic valve, Loeys- Dietz syndrome, hereditary thoracic aortic aneurysm or vascular Ehlers- Danlos syndrome.1 Congenital diseases eg coarctation of aorta, Turner syndrome or tetralogy of Fallot also predispose to dissection in young persons. Risk of type A dissection is greatest when the aortic root diameter is more than 40 mm.1 Patients with dissection involving ascending aorta have higher risk of subsequent rupture, pericardial effusion, aortic insufficiency and coronary involvement. Aortic regurgitation in the setting of type A dissection can be due to several mechanisms. First, there may be incomplete coaptation of aortic leaflets because of associated dilatation of aortic root and annulus.2 Second, dissection may lead to prolapse of aortic leaflet. Third, circumferencial dissecting intimal flap may itself prolapse into the left ventricular outflow tract during diastole and cause severe aortic regurgitation.1 Accurate diagnosis is essential. Transesophageal echocardiography is at least equal to or even superior to CT and aortography in the diagnosis of aortic dissection.3 Proximal aortic dissection usually requires immediate surgical repair.

References

1. Braverman AC, Thompson RW, Sanchez LA. Diseases of the Aorta. In Bonow RO, Mann DL,
