Left main trunk connecting to superior vena cava via aneurysmal coronary artery fistula

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A 3-year-old male child presented with an incidentally detected continuous murmur during evaluation for respiratory tract infection. Two-dimensional echocardiography (Panel A) showed an aneurysmally dilated left main coronary artery (CA) with the presence of large coronary cameral fistula originating from the left main CA. Coronary computed tomography (Panels B and C) showed CA fistula extending from the left main CA, and distally opening into right-sided superior vena cava (SVC), proximal to superior vena cava-right atrium (SVC-RA) junction. The fistulous communication is seen as a tortuous vessel coursing posterior to aorta in the interatrial groove. The maximum diameter was 12 mm. CA fistulas are reported in 0.1–0.2% of all patients undergoing selective coronary angiography. The major sites of origin of fistulae are right coronary artery (35%), left coronary artery (35%), and both coronary arteries. The major sites of terminations are right ventricle (40%), right atrium (26%), pulmonary artery (17%) less frequently SVC, CS and least in left atrium and left ventricle (2%).

We report this case as proximal fistulae arising from the left main CA are rare and so also their opening in SVC is a rare entity.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.

Reference