Comment

Progressive VSD restriction in DORV with remote VSD is rare but should deserve special attention in the patient's follow-up. To our knowledge, there are two reports describing patients with DORV and two well-formed ventricles who became critically ill as a result of the closing VSD [1, 2]. Both died, unfortunately. Our patient was successfully managed to reach biventricular repair after opening and enlargement of the closed perimembranous VSD that became apparent by LV hypertension caused by the closing muscular VSD. We believe that the surgical opening of the VSD in the perimembranous area was a crucial step for the subsequent biventricular repair in terms of the recovery of LV function and the creation of an LV outlet near the aortic valve.

Treatment strategy for patients with DORV and a restrictive VSD depends on the size of the LV and the location of the VSD. Some patients are amenable to biventricular repair, whereas others are subjected to univentricular palliation. Patients with good-sized and well-functioning ventricles and an enlargeable VSD near an outlet valve are good candidates for biventricular repair. Serraf and colleagues [3] reported two early deaths attributable to LV hypertrophy in 30 patients with DORV and a restrictive VSD undergoing biventricular repair. The staged approach interposing palliative LV decompression may contribute to further reduction in the mortality.

Reports describing transcatheter VSD creation and enlargement in patients with DORV and a severely restrictive VSD [4, 5] asserted the following merits of the transcatheter approach: patients in an unstable condition could avert general risks associated with surgical interventions, and damage to the aortoventricular valves or conduction system is prevented by targeting the midmuscular portion remote from these structures. However, aortoventricular valve regurgitation and myocardial perforation had been reported as complications of the transcatheter approach. Recurrent stenosis as a result of VSD stent obstructions is common [5]. We believe that surgical VSD enlargement is preferable to the transcatheter approach in patients who are amenable to biventricular repair, because a midmuscular VSD created by the transcatheter approach is not suitable for intraventricular rerouting.

References


Congenital Aneurysm of the Aortmitral Intervalvular Fibrosa

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A 5-year-old boy was found to have a congenital left ventricular outflow tract (LVOT) aneurysm of the inter-
vallular fibrosa, LVOT obstruction after repair of a per-
imembranous ventricular septal defect, and aortic coarctation. The patient underwent successful plication of the aneurysm, resection of the fibrous subaortic stenosis, and septal myectomy.  


Aneurysms of the aortermitral curtain or the inter-
vallular fibrosa are rare and have only been 
described previously in association with infective endo-
carditis [1, 2] or postsurgical trauma [3]. We report a case of congenital aneurysm of the intervalvular fibrosa presenting as a left ventricular outflow tract (LVOT) aneurysm in association with LVOT obstruction. The

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Children’s National Medical Center approved a waiver of informed consent for this retrospective study.

A 5-year-old boy presented with increasing LVOT obstruction. He had previously undergone repair of a perimembranous ventricular septal defect and aortic coarctation at 2 months of age. At 4 years of follow-up, a transthoracic echocardiogram and a cardiac magnetic resonance image revealed moderate subaortic stenosis (peak gradient of 86 mm Hg) with flow acceleration beginning at the crest of the ventricular septum. Distal to the area of obstruction, a saccular 4 × 4 cm complex LVOT aneurysm was noted extending up to the aortic valve annulus. The mass projected toward the interatrial septum, indenting toward the right atrium (Fig 1).

Using a median sternotomy and cardiopulmonary bypass, the subaortic area was approached through a transverse ascending aortotomy and a right atriotomy to confirm the relationship of the aneurysm to the aortic and mitral valves as well as the interatrial septum. A 4 × 4 cm saccular dilatation was noted between the aortic and mitral valves (in the region of the aortomitral intervalvular fibrosa) (Fig 2A). The aneurysm was clear of any thrombus. The mitral valve was normal in appearance. Additional complex subaortic stenosis resulting from a fibromuscular membrane was noted extending to the

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**Fig 1.** Preoperative (A) transesophageal echocardiography and (B) cardiac magnetic resonance imaging showed large 4 × 4 cm aneurysm of aortomitral intervalvular fibrosa (small arrows) and bowing of interatrial septum into the right atrium. Large arrows denote the position of the aortic valve annulus, and arrowheads denote the level of left ventricular outflow tract (LVOT) obstruction resulting from fibromuscular ridge.

**Fig 2.** Intraoperative images. (A) Transverse ascending aortotomy showing saccular aneurysm in aortomitral intervalvular fibrosa (arrowhead), left ventricular outflow tract (LVOT) (large arrow), and aortic valve (small arrow). (B) Sutures were placed through the right atrium and through the interatrial septum at the lower margin of the aneurysm (white arrows) into the LVOT, picking up multiple bites through the sac of the aneurysm, and were passed back into the right atrium through the superior margin of the aneurysm (black arrows).

**Fig 3.** Postrepair transesophageal echocardiography. (A) Two-dimensional and (B) color Doppler showing laminar flow across the left ventricular outflow tract (LVOT) with no residual aneurysm; plicated aneurysm wall (large arrow), LVOT (arrowhead), and aortic valve (small arrow).
anterior leaflet of the mitral valve upstream of the aneurysm.

After excision of the obstructing fibromuscular membrane and a standard septal myectomy, the aneurysm was plicated using pledgeted horizontal mattress sutures of 4-0 Prolene (Ethicon, Somerville, NJ). The sutures were placed through the right atrium and through the interatrial septum at the lower margin of the aneurysm into the LVOT, picking up multiple bites through the sac of the aneurysm, and was passed back into the right atrium through the superior margin of the aneurysm. This allowed the pledgets to be seated in the right atrium to minimize pledget load and scarring in the LVOT (Fig 2B).

A postrepair transesophageal echocardiogram demonstrated laminar flow across a normal-appearing LVOT and no residual aneurysm. Both the aortic and mitral valves were competent (Fig 3). The patient had an uneventful recovery, was discharged home 5 days postoperatively, and continues to do well without any residual hemodynamic lesions at 6-month follow-up.

Comment

Aneurysms of the aortomitral curtain or the intervalvular fibrosa are rare and have only been described previously in association with infective endocarditis [1, 2]. To our knowledge, this is the first description of congenital aneurysm of aortomitral intervalvular fibrosa.

Our patient had undergone a ventricular septal defect and aortic coarctation repair in the past. On retrospective review of the patient’s imaging studies before the procedure, a smaller dilatation was noted in the region of the aortomitral intervalvular fibrosa, which subsequently progressed to its current size. Although pseudoaneurysms of the LVOT have been reported with injury to the area from surgical procedures [3], it was unlikely in this patient, because the findings preceded the first surgical intervention.

Because these aneurysms per se were asymptomatic, clinical detection of this pathologic process depended on the presence of other associated symptomatic lesions. Although the natural history of such lesions is not well known, surgical repair of aortomitral intervalvular fibrosa is justified based on the progressive enlargement of the aneurysm, as seen in our case.

References


Histologic Evolution From Adenocarcinoma to Squamous Cell Carcinoma After Gefitinib Treatment

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We report two cases of lung cancer with histologic transformation from adenocarcinoma to squamous cell carcinoma after gefitinib treatment. Both cases involved advanced lung cancers, initially confirmed as adenocarcinomas with sensitive epidermal growth factor gene mutations. After gefitinib treatment, the second pathologic examination in each case revealed squamous cell carcinoma retaining identical mutations without newly acquired resistance mutations. The underlying mechanism may have been pluripotent tumor cells with divergent differentiation or mixed lung cancer including both adenocarcinomatous and squamous cell carcinomatous components. This report widens the spectrum of histologic evolution as a mechanism underlying the acquisition of drug resistance.


Gefitinib and erlotinib, selective tyrosine kinase inhibitors (TKI) of epidermal growth factor receptor (EGFR), have demonstrated effectiveness against adenocarcinomas with EGFR mutation. Herein, we report two cases of biopsy-confirmed lung adenocarcinomas that underwent histologic evolution to squamous cell carcinoma (SCC) after gefitinib treatment.

Case Reports

Case 1

A 51-year-old woman presented with symptoms of coughing and a loss in body weight of 7 to 8 kg during a period of 3 to 4 months. Roentgenogram and computed tomography scans revealed a mass greater than 8 cm in the right lung (Fig 1A) accompanied by multiple pleural nodules. Video-assisted thoracoscopic surgery confirmed pleural metastasis, and pathologic examination revealed a metastatic adenocarcinoma with intracytoplasmic mucin and glandular structures (Fig 1B). Immunohistochemical analysis confirmed primary lung

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