peak-to-peak gradient). Re-recoarctation occurred, necessitating reoperation at 15 months. The aortic arch was enlarged with a bovine patch (Cardiofix pericardium; Carbomedics, Inc, Arvada, Colo). Follow-up for 3½ years demonstrated no residual aortic obstruction.

Patient 3

A 2½-month-old baby boy weighing 4 kg had COA. Echocardiography showed discrete isthmic COA, closed ductus arteriosus, bicuspid aortic valve, and normal pulmonary branches. FISH analysis performed because of hypercalcemia confirmed WBS. Surgery comprised an end-to-end anastomosis. Complete resection of all pathologic tissue was not feasible because the narrowing of the descending aorta extended too far distally. Postoperatively, mild COA persisted with a systolic gradient of 25 mm Hg. Reoperation at 6 months was performed with patch enlargement of the thoracic descending aorta. Follow-up for 3 years demonstrated no residual aortic obstruction.

DISCUSSION

Neonatal diagnosis of WBS is difficult because the syndrome usually occurs sporadically and the striking physical and behavioral features are rarely recognizable. Rather, patients typically present after 1 year of age.[1,2] Moreover, conversely to older children in whom supravalvular aortic stenosis and/or peripheral pulmonary stenosis are often associated,[1] COA is often isolated in infants with WBS.[2-5] Finally, although COA in infantile WBS may take the atypical form with extension far beyond the isthmus in the so-called middle aortic syndrome,[5] it rather mimics a typical neonatal COA, as in our 3 cases. However, infantile COA in the setting of WBS is not exceptionally encountered. In a recent study focusing on 129 cases of infantile WBS, COA was the fourth more frequent cardiovascular lesion, in 19% of cases.[2] Our case series suggests that WBS might be encountered in approximately 2% of cases pf neonatal or infantile COA.

Recoarctation occurred in our 3 patients after initial surgery, similar to the 3 previously published cases of WBS with neonatal COA.[3-5] Transcatheter balloon dilation seems disappointing to treat recoarctation in WBS, and stent implantation is not appropriate in small infants. The appropriate treatment is probably surgery with patch enlargement and even sometimes exclusion of the stenotic aortic segment.

We acknowledge Michel Voisin, MD, from Hôpital Arnaud de Villeneuve, Montpellier, France, for patient referral, and Phalla Ou, MD, PhD, from Hôpital Necker-Enfants-Malades, Paris, France, for providing computed tomography images.

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Aortic dissection caused by aortitis associated with hepatitis C virus–related cryoglobulinemia

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Disclosures: None.
Received for publication June 29, 2010; accepted for publication July 11, 2010; available ahead of print Aug 23, 2010.
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J Thorac Cardiovasc Surg 2010;140:e81-2
0022-5223/$36.00

doi:10.1016/j.jtcvs.2010.07.021

Cryoglobulinemia can generally cause systemic vasculitis to small and medium-sized vessels, not to great vessels, resulting in neuropathy, renal impairment, and coagulopathy. To our knowledge, this is the first description of acute aortic dissection caused by aortitis associated with hepatitis C virus–related cryoglobulinemia.

CLINICAL SUMMARY

A 71-year-old man was referred for emergency surgery for acute type A aortic dissection in an intubated state. He

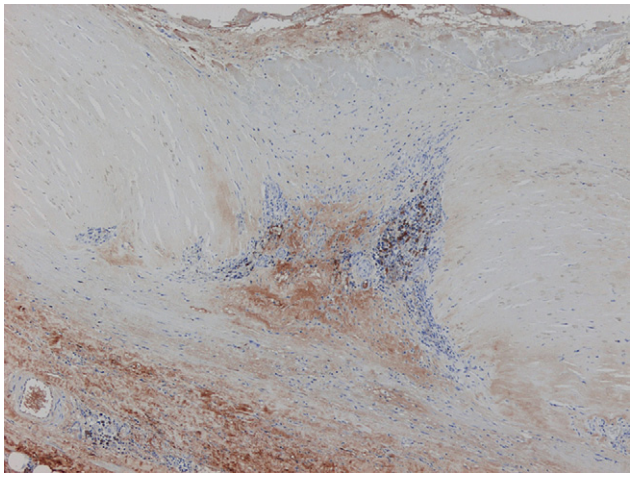

FIGURE 1. Immunofluorescence of aortic wall shows the presence of immunoglobulin G.

had taken 10 mg of oral prednisolone for vasculitic neuropathy induced by hepatitis C virus–related cryoglobulinemia for about 4 years.

On arrival, blood pressure was stable and results of laboratory examinations were unremarkable. Physical examinations showed a regular heart rate without any murmur. Enhanced computed tomography showed type A aortic dissection of the ascending aorta without pericardial effusion. Transthoracic echocardiography revealed no evidence of aortic regurgitation. Emergency hemiarch reconstruction with a prosthetic graft was performed under deep hypothermic circulatory arrest and retrograde cerebral perfusion, followed by antegrade cerebral perfusion on cardiopulmonary bypass.

He was extubated 75 hours after the operation and weaned from catecholamine with ease in the intensive care unit. During intubation, intravenous steroid was administered. Subsequently, we initiated 10 mg of prednisolone again. His clinical course was uneventful and he was discharged on foot without any complications.

The histopathologic finding of aortic wall showed the chronic infiltration of inflammatory cells such as lymphocytes and plasma cells within the media and around the vaso vasorum, suggestive of aortitis. Immunofluorescence revealed immunoglobulin G in the aortic wall (Figure 1).

DISCUSSION

Aortitis is typically a chronic, progressive disease associated with large vessels, in particular the ascending aorta. The main causes of aortitis are classified into 2 categories: infectious or noninfectious. The former includes syphilis, *Salmonella, Staphylococcus,* and other bacteria. The latter includes some forms of primary or secondary vasculitis. Giant cell arteritis and Takayasu arteritis are the most common causes of noninfectious aortitis. Others are relapsing polychondritis, Cogan syndrome, sarcoidosis, rheumatoid arthritis, systemic lupus erythematosus, and spondyloarthropathies.[1,2] However, these other systemic diseases are rarely associated with aortitis. Only 5 cases of aortic dissection associated with aortitis were recognized, according to the study by Rojo-Leyva et al.[2]

Cryoglobulinemia with abnormal immunoglobulin precipitates appearing at a temperature of 4°C has clinical manifestations including vasculitis, neuropathy, renal impairment, and coagulopathy. These have been induced by cryoprecipitable circulating immune complex. In general, vasculitis of small and medium-sized vessels has been widely recognized. Additionally, in the absence of any symptoms, many organs with vessels of these sizes may be involved. However, great vessel vasculitis has not been reported in the English medical literature.

Au and associates[3] reported an association between cryoglobulinemia and structural aortic abnormality. They presented a case of aortic dissection in which aortitis was not confirmed on histopathologic findings. Additionally, they proposed the mechanism causing aortic aneurysm or dissection, which was the intimal damage to great vessels concomitant with development of cryoglobulins.

The deposition of immune complex in vessel walls gives rise to systemic vasculitis and various associated symptoms.[4] Immnofluorescence study shows the presence of immune reactants such as immunoglobulins G, M, and/or C3.

In our case, the infiltration of inflammatory cells and the deposition of immunoglobulin G were confirmed in the aortic wall in the context of hepatitis C virus–related cryoglobulinemia. The cause of aortic dissection could be accounted for by aortitis associated with these mechanisms. To our knowledge, this is the first description of acute aortic dissection caused by aortitis associated with hepatitis C virus–related cryoglobulinemia.

We are grateful to Dr D Yamashita, Dr H Arima, and Dr Y Imai for their special advice.

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