Hemothorax and Hemopericardium in a Patient with Bean's Blue Rubber Bleb Nevus Syndrome*

David Langleben, M.D.;† Norman Wolkove M.D., F.C.C.P.; Herbert Srolovitz, M.D.; Robin C. Billieck, M.D.; and Nathan M. Sheiner, M.D.

A patient with Bean's blue rubber bleb nevus syndrome presented with a hemothorax, and 18 years earlier had presented with "idiopathic" hemopericardium and tamponade. Typical hemangiomas were found in the pleura and skin. This is the first report of intrathoracic bleeding with this disorder. (Chest 1999; 95:1352-53)

In 1958, Bean described rubbery blue hemangiomas of the skin and gastrointestinal tract associated with gastrointestinal bleeding, which he termed the "blue rubber bleb nevus syndrome." Subsequent papers have identified hemangiomas in other visceral organs, including the pleura. Pleural bleeding, however, has not been reported. We now describe a patient who presented with a hemothorax and had the classic skin findings of blue rubber bleb nevus syndrome and hemangiomas of the pleura. This patient had also developed a hemopericardium 18 years prior to the hemothorax, possibly related to the same disease process.

CASE REPORT

The patient is a 58-year-old retired textile cutter of Italian origin who smokes cigarettes and denies asbestos exposure. In 1968, he had presented to another hospital in cardiac tamponade, with a two-week history of orthopnea and leg edema. The chest roentgenogram revealed an enlarged globular cardiopericardial silhouette and a density at the left base suggestive of effusion. Pericardiocentesis removed 1,100 ml of serosanguinous fluid. The tamponade recurred, requiring another pericardiocentesis. A left thoracocentesis extracted 700 ml of serosanguinous fluid containing fibrin, hemolyzed red cells, lymphocytes, and mesothelial cells. Cultures of both fluids were negative. There was no clinical or laboratory evidence for malignancy, tuberculosis, or autoimmune disease. Acute and convalescent viral titers for coxsackie A9 were 1/24 and 1/768, and for coxsackie B4 1/48 and 1/64. The patient was treated with prednisone and then continued on diuretics for the next 18 years. He was asymptomatic and working full time.

In March 1986, he presented to another hospital with a one-month history of dyspnea and a right sided pleural effusion. Thoracocentesis extracted bloody fluid which did not contain malignant cells or Mycobacteria and which increased despite furosemide therapy.

In July 1986, he was seen at our hospital where vital signs were normal. The chest roentgenogram showed right-sided pleural fluid and marked pleural thickening. Thoracocentesis removed serosanguineous fluid containing only blood cells. A right thoracotomy was performed, 1,000 ml of bloody fluid was removed, and the right lung was found to be surrounded by a thickened fibrous parietal pleura and was trapped within a similarly thickened visceral pleura. A pleural and superficial lung biopsy showed vascular lesions (capillary and arteriolar-venular type), arranged in a lobular pattern, supported by a loose connective tissue stroma attached to the visceral pleura with a short stalk (Fig 1). The endothelial cells were either flat or slightly swollen, but possessed no atypical cytologic features. Except for slight fibrotic thickening of the pleura, the pleura and lung were otherwise unremarkable. Granulomas and malignant cells were absent. No specific therapy was administered and ankle swelling has persisted despite furosemide therapy. The pleural effusion has not recurred and a recent cardiac catheterization suggested mild pericardial constriction.

On a recent visit, multiple violaceous compressible venous lakes were recognized over the arms, chest, and back, and the patient said these were a lifelong finding. The diagnosis of blue rubber bleb nevus syndrome was considered and a skin biopsy was performed. It showed an attenuated epidermis in which a cavernous hemangioma was present (Fig 2). The vascular walls were thin, endothelial cells typical, and several fibrin thrombi were present.

DISCUSSION

The term "blue-rubber-bleb-nevus," first coined by Bean, applies to a syndrome of rubbery angiomas of the skin, variable in size and number which are compressible and refill upon release of the compression. Associated with this cutaneous abnormality, angiomas of the Gl tract have been reported to cause profuse bleeding. Although rare, the syndrome has been recognized in the literature and over 40 studies have been reported.

![Figure 1: Lobular capillary-arteriolar-venular hemangioma with loose connective tissue stroma attached to a thickened pleural surface (bottom of figure) (hematoxylin-eosin, original magnification X 100).](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21595/...)

*From the Divisions of Cardiology, Pulmonary Diseases, and Dermatology, Department of Medicine, and Departments of Pathology and Surgery, Sir Mortimer B. Davis Jewish General Hospital, and McGill University, Montreal, Canada.
†Scholar of the Canadian Heart Foundation.

Reprint requests: Dr. Langleben, Hotel Juif, 3755 Cote Ste Catherine, Montreal, Quebec, Canada H3T 1E2

1352

Bean's Blue Rubber Bleb Nevus Syndrome (Langleben et al)
cases reported. Tissues and organs affected include the liver, spleen, central nervous system, skeletal muscle, heart, oro- and nasopharynx, adrenal glands, kidney, thyroid, and glans penis. Pleural and lung involvement has been mentioned, but pleural hemorrhage has not previously been described.

Our patient presented with a hemothorax, had pleural angiomas detected by histologic study, and had typical skin findings. Infectious and autoimmune etiologies for the pleural effusion were excluded. It is likely that the bleeding angioma caused the pleural effusion.

We do not know if our patient has pericardial involvement, although angiomas of the pericardium have been described in the blue rubber bleb nevus syndrome. The presence of angiomas in the pericardium might predispose to bleeding spontaneously or from a viral pericarditis. In this regard, it is interesting that our patient had a significant rise in titers for coxsackie A9 virus. However, this agent is not known to affect either pericardium or pleura. More likely, therefore, the pericardial bleed represented a spontaneous event in a patient with pericardial angiomas. Aside from gastrointestinal, nasal, and genitourinary bleeding, hemorrhage from other sites has not previously been described in the blue rubber bleb nevus syndrome. We have shown that intrathoracic bleeding can occur in this syndrome and may pose life-threatening problems. Although the syndrome is uncommon, it may often be recognized by a careful skin examination and should be familiar to all internists who diagnose and manage pleural and pericardial effusions.

ACKNOWLEDGMENTS: We thank Christina Tsioutsias for preparation of the manuscript.

REFERENCES

6 Wood MW, White RJ, Kernohan JW. Cancerous hemangiomas involving the brain, spinal cord, heart, skin and kidney. Staff Meetings of Mayo Clinic 1957; 32:249-54
9 Smart RH, Newton DE. Hemangiomata of the penis with blue rubber bleb syndrome. J Urol 1975; 113:570-71

Reinforcement of the Anastomosis between the Fragile Arterial Vessel and the Prosthetic Graft*

A Headband Method

Kiyoshi Ishii, M.D.; Yasunori Koga, M.D.; Kaoru Isamoto, M.D.; Yoshio Onitsuka, M.D.; and Kouichiro Shibata, M.D.

A new method of anastomosis was devised, intended for the reinforcement of the anastomosed area between the fragile vessel of the patient and the prosthetic graft in cardiovascular surgery. This method, which may be called a headband method, consists of suturing a small prosthetic graft wrapping the anastomosed area together with the arterial vessel in the manner of a headband. Fourteen cases have been evaluated at follow-up periods ranging from 24 to 40 months. All cases proved successful without noteworthy complications. (Chest 1989; 95:1353-54)

The fragility of arterial vessel walls in dissecting aneurysms or other aneurysms adds a difficulty to the success of anastomosis of arterial vessels and prosthetic grafts. In

*From the Second Department of Surgery, Miyazaki Medical College, Miyazaki, Japan.
Reprint requests: Dr. Ishii, 2nd Department of Surgery, Miyazaki Medical College, 5200 Ichara Kayotake-cho, Miyazaki Prefecture 889-16, Japan

Figure 2. Cavernous hemangioma expanding the papillary and reticular dermis. Two fibrin thrombi are present (hematoxylin-eosin, original magnification x 100).