or venous route, gravely affect clinical outcomes. In this respect, distant metastasis of invasive thymoma via such routes is considered rare, amounting to a few percent in primary cases and to a maximum 26 percent in recurrent cases, indicating that treatment of the local lesions related to thymoma may be the most pressing problem.

Treatment of disseminated lesions of invasive thymoma in the pleural and/or pericardial cavity has included chemotherapy (including immunotherapy), radiotherapy, surgery, and their various combinations. Chemotherapy agents, including cisplatin, cyclophosphamide, doxorubicin, vincristine, and corticosteroids are commonly applied, and partial to unilateral irradiation of the total thoracic field at varying doses has been performed. Surgical approaches have included partial resection of the lung and/or pleura to the greatest extent, and, when possible, panpleuropneumonectomy. However, as disseminated lesions often show varying degrees of resistance to such therapies, repeated relapse during the clinical course may lead to death. Intrathoracic chemothermotherapy combined with surgical therapy may represent a new treatment modality for residual disseminated lesions of invasive thymoma which addresses this problem.

This patient showed little response to chemotherapy and radiotherapy at another hospital, and even panpleuropneumonectomy might have been insufficiently radical, because of the large number of widely disseminated lesions in the pleural cavity. Cytologic examination following panpleuropneumonectomy, in fact, demonstrated that the pleural effusion contained malignant cells. However, after the third course of intrathoracic chemothermotherapy, the cytologic results for the pleural effusion became negative; this finding was confirmed at the subsequent exploratory operation performed because of empyema, and the patient is now free from tumor. Considering these observations together, the intrathoracic chemothermotherapy in the present patient might be an apparently extremely effective postoperative adjuvant therapy.

At present, as the effect of intrathoracic chemothermotherapy on thymoma has not been sufficiently analyzed, for example, using in vitro assay to study chemothermotheral sensitivity of thymoma cells, it is unknown whether this therapy may be routinely applied to malignant pleurisy of invasive thymoma; this issue requires further study. Nevertheless, on the basis of our clinical experience in the present case, we are hopeful that intrathoracic chemothermotherapy may become a useful adjuvant treatment to control disseminated lesions in the pleural cavity, especially when other therapies are unsuccessful or insufficient.

REFERENCES

Aortic Valve Papillary Fibroelastoma*  
A Diagnosis by Transthoracic Echocardiography

Aasha Gopal, M.D.; Giuseppe Li Mandri, M.D., Ph.D.; Donald L. King, M.D.; Charles Markov, M.D.; and Shunichi Homma, M.D., F.C.C.P.*

*From the Department of Medicine, Divisions of Cardiology (Dr. Gopal, Li Mandri, King, and Homma) and Pathology (Dr. Markov), Columbia-Presbyterian Medical Center, New York. Reprint requests: Dr. Homma, Cardiology Division, Columbia-Presbyterian Medical Center, New York, NY 10032.

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Cardiac papillary fibroelastomas are unusual, frond-like growths typically found on cardiac valves, diagnosed incidentally on autopsy or cardiac surgery, but rarely during life. We report a rare case of an aortic valve papillary fibroelastoma detected by transthoracic echocardiography and confirmed by histologic study.

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Papillary fibroelastomas are benign lesions that account for less than 1 percent of all primary cardiac tumors and represent the majority of valve tumors.\(^{1,2}\) They have been detected incidentally during cardiac surgery or autopsy and have been described in all age groups and both sexes, although mostly in adults over 60 years. The occasional detection of papillary fibroelastomas during life has been made possible by the increasing use of transthoracic echocardiography.

CASE REPORT

A 49-year-old man was referred for evaluation of lightheadedness, chest tightness, and an episode of breathlessness 1 month prior to hospital admission. Results of physical examination, laboratory data, blood cultures, chest radiography, and ECG were unremarkable. Thallium stress testing and Holter monitoring were also normal.

 Transthoracic echocardiography revealed normal left ventricular function and a 1.4 X 1.5-cm mobile mass attached to the aortic aspect of the noncoronary cusp of the aortic valve (Fig 1). There was no evidence of aortic stenosis or regurgitation. Because of the proximity of the lesion to the coronary ostia and the concern of embolic risk, cardiac catheterization was deferred and surgical removal of the tumor was recommended.

At surgery it was found that the aortic valve was tricuspid and presented a soft papillary lesion arising from the edge of the noncoronary aortic cusp. The tumor was excised completely together with a small edge of the aortic leaflet. As the tumor excision was possible through the aorta, a ventriculotomy was not required. On pathologic examination the tumor appeared as a tan-colored, 0.5 X 0.05 X 0.3-cm pedunculated lesion with numerous papillary fronds arising from a central stalk. The diagnosis of papillary fibroelastoma was confirmed on microscopic examination (Fig 2).

 Transthoracic echocardiography 1 week after the operation revealed normal aortic valve motion with only trace aortic regurgitation by color flow Doppler interrogation. The patient recovered well and has since been asymptomatic.

DISCUSSION

Papillary fibroelastomas are benign tumors found on the surface of papillary muscle, chordae tendineae, ventricular septum, and endocardium of all cardiac chambers and they may occur on either side of cardiac valves. Pathologically, these tumors appear as multiple papillary villous fronds radiating from a dense central stalk. The etiology of these lesions remains unclear, but it is thought that they may derive from the endocardium itself.\(^{2}\)

Because the tumors are small (usually <1 cm in diameter), they are largely asymptomatic. Occasionally, they may give rise to emboli leading to transient ischemic attacks and cerebral infarction.\(^{3}\) Aortic valve papillary fibroelastomas have also been reported to cause intermittent occlusion of the coronary ostia giving rise to angina, myocardial infarction, or sudden death.\(^{4,5}\)

Although in autopsy series aortic valve lesions seem to be more common than mitral valve papillary fibroelastoma, diagnosis of these aortic valve tumors during life is very unusual and has been reported rarely.\(^{1}\) To our knowledge, our case represents only the third antemortem transthoracic echocardiographic diagnosis of a papillary fibroelastoma on the aortic valve.\(^{5,6}\) It is unclear whether our patient’s symptoms were coincidental or directly related to the tumor.

 Transthoracic echocardiography, occasionally complemented by cardiac magnetic resonance imaging and transesophageal echocardiography, is very useful in the detection of these small intracardiac lesions. There are no definitive echocardiographic features, however, that distinguish papillary fibroelastoma from other intracardiac masses.

Though most fibroelastomas are asymptomatic, they carry a significant embolic risk and therefore surgical resection is usually recommended. In most cases, simple tumor excision is sufficient, but extensive valvular involvement may require valve repair or replacement.\(^{3,4}\) As the dimensions of the tumor can be assessed relative to that of the left ventricular outflow tract, the feasibility of tumor excision through the aortic orifice without subjecting the patient to the risk of ventriculotomy can be determined by echocardiography.

In conclusion, echocardiography is very important in both detecting and planning the surgical approach to papillary fibroelastoma. Transthoracic echocardiography should also be considered in the investigation of strokes and
angina of unclear etiology as they may result from the embolization of these unusual tumors.

REFERENCES

Bronchogenic Squamous Cell Carcinoma Complicating Localized Recurrent Respiratory Papillomatosis*

Eric Wilde, M.D.; Maitre A. Duggan, M.B.; and Stephen K. Field, M.D.

Bronchogenic squamous cell carcinoma has been reported in patients with recurrent respiratory papillomatosis (RRP) extending into the tracheobronchial tree even in the absence of a history of radiation therapy or smoking. We present a case of bronchogenic squamous cell carcinoma developing in a patient with RRP localized to the larynx for 45 years. (Chest 1994; 105:1887-58)

| HPV=human papilloma virus; RRP=recurrent respiratory papillomatosis |

Recurrence respiratory papillomatosis (RRP) is the most common benign respiratory tract neoplasm in childhood. It is usually limited to the larynx but can extend into the tracheobronchial tree and par enchyma.

Recurrent respiratory papillomatosis is associated with human papilloma virus (HPV). More than 66 types of HPV have been identified by DNA hybridization techniques. Human papilloma viruses 6 and 11 are associated with the majority of RRP cases.

Squamous cell carcinoma of the larynx and tracheobronchial tree is an unusual complication of RRP. Most cases of squamous cell carcinoma complicating RRP have occurred in smokers or patients treated with irradiation. However, some cases have recently been reported in patients without these risk factors.

Previously reported cases of bronchogenic carcinoma

*From the Foothills Hospital and University of Calgary, Calgary, Alberta, Canada.

Figure 1. Most of the nuclei in this nest of squamous carcinoma contain stainable HPV 11 DNA (in situ hybridization with biotinylated probes to HPV 11 and counterstained with nuclear fast red, original magnification X200).

have been in patients with RRP extending into the tracheobronchial tree or par enchyma. We report a case of bronchogenic squamous cell carcinoma where the RRP was clinically localized to the larynx.

CASE REPORT

A 48-year-old woman was originally diagnosed as having RRP at 2 years of age when she presented with hoarseness and dyspnea. She had six separate tracheotomies for laryngeal papilloma resection in childhood. Following these, about 18 endoscopic laryngeal papilloma resections were performed. Between 1986 and July 1991, five endoscopic laser resections were performed. After the last procedure, she developed a persistent cough. In November 1991, she developed fever and purulent sputum. She denied hemoptysis but had lost 6 kg over the preceding 4 months. She was never irradiated and is a lifelong nonsmoker. Breath sounds were diminished at the right base and the patient was hoarse. Results of the physical examination were otherwise unremarkable.

A chest radiograph showed a right lower lobe infiltrate. No other abnormality was noted. A course of erythromycin therapy improved the fever and sputum but the cough persisted. A repeated chest radiograph was unchanged. A computed tomographic scan demonstrated occlusion of the superior segment bronchus with distal consolidation.

During bronchoscopy, five papillomata were seen in a distorted larynx. The trachea and left bronchi cal tree were unremarkable. Concentric narrowing and virtual occlusion of the right superior segment bronchus were seen. No papillomata were seen. Bronchial washings demonstrated malignant squamous cells.

The patient underwent a right pneumonectomy and removal of a subglottic papilloma. The lower lobe was almost totally replaced by a 9 x 8 x 6.5-cm firm, partially necrotic mass. The mass extended into the bronchial lumen where it had a papillary growth pattern. Histologically, the tumor was mostly solid with a papillary and solid architecture. Cavitation due to necrosis was a prominent feature. The papillae and solid nests were formed of large polygon epithelial cells that displayed prominent keratination toward the center. Cellular pleomorphism and mitotic activity were easily identified. The tumor was an invasive, moderately differentiated squamous cell carcinoma that had replaced

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