Giant Right Atrial Thrombus in Noonan Syndrome Combined with Eisenmenger's Complex

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A 54-year-old woman with the Noonan syndrome was admitted with congestive heart failure and a giant right atrial thrombus with atrial septal defect detected by two-dimensional echocardiography. The thrombus vanished on oral anticoagulant therapy with warfarin. The thrombus is considered to result from hemostasis in the right atrium due to congestive heart failure and to her specific skeletal characteristics. This report describes the first case of Noonan syndrome with right atrial thrombus.

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Through advances in two-dimensional echocardiography, intracardiac thrombus, which could not be diagnosed previously can now be detected more readily. There have been many reports of thrombus of the left side of the heart, but intracavitary thrombus of the right side is rare and only a few antemortem cases of right atrial thrombus are reported. We present a patient with giant right atrial thrombus detected by two-dimensional echocardiography in the presence of Noonan syndrome combined with Eisenmenger's complex.

Case Report

A 54-year-old woman was admitted to our hospital for treatment of dyspnea. Cardiac catheterization, performed at the age of 40, revealed atrial septal defect, which advanced to Eisenmenger's complex. Since then, she had repeated attacks of congestive heart failure. She was 128 cm high, and had mental retardation and facial and skeletal abnormalities (Fig 1). Chromosomes were normal. From these findings, she was considered to have Noonan syndrome.

Physical examination on admission revealed that the patient had an irregular pulse rate, and blood pressure of 98/60 mm Hg. The second heart sound was split widely and fixed. Systolic ejection murmur (3/6) and mid-diastolic rumbling murmur (3/6) were auscultated. The jugular veins were moderately dilated with mild hepatomegaly and peripheral edema, but there was no clinical evidence of deep vein thrombosis. The chest x-ray film represented marked cardiomegaly with congestion in bilateral lung fields. Computed tomography of the chest revealed marked enlargement of bilateral atra and a giant round mass in the diverticular segment of the right atrium. An electrocardiogram showed atrial fibrillation and right ventricular hypertrophy. There was no abnormality in blood coagulation.

Two-dimensional echocardiography showed marked right and left atrial dilatation and a large interatrial septal defect. A giant round homogenous mass was demonstrated consistently in the right atrium. The mass adhered to the right atrial free wall and was immobile. A "smoke-like" echo was noted around the mass (Fig 2, left). From these findings, this mass was diagnosed as a right atrial thrombus. With management for congestive heart failure, anticoagulant therapy with warfarin was started after detection of the thrombus, because the patient rejected operation. The thrombus was decreased in size to half after 40 days of anticoagulant therapy (Fig 2, middle). It was no longer detected three months later by two-dimensional echocardiography, but a "smoke-like" echo remained in the right atrium (Fig 2, right). No clinical symptom or sign of pulmonary or systemic thromboembolism was observed during the three-month period of thrombus resolution.

FIGURE 1. A 54-year-old, 128-cm woman with Noonan syndrome. The skeletal deformities include webbing of the neck, scoliosis, and pectus carinatum. The facial abnormalities consist of epicanthal folds, ptosis of eyelids, hypertelorism, strabismus, and low-set ears.
**DISCUSSION**

Wartman and Hellerstein\(^1\) reported in their review of more than 2,000 autopsies that thromboemboli of the right atrium and ventricle were found in only 14 cases. Moreover, antemortem diagnosis has been made only in a few cases.\(^2,3\)

An antemortem case without endocardial injury or intracardiac equipment such as pacemaker leads and hyperalimentation catheters is uncommon. From previous echocardiographic\(^4\) and postmortem reports, many cases of right atrial thromboemboli revealed emboli from deep vein thrombus. These are long in shape, extremely mobile, prolapsing across the tricuspid orifice, and pulmonary arterial embolization is frequently noted.\(^5,6\) On the contrary, right atrial thrombus that develops in situ is shorter, round, immobile and adheres to the lower lateral right atrial wall by broad-based stalk. Felner et al.\(^7\) listed ten of the reported cases of nonprolapsing right atrial thromboemboli that developed in situ, forming on a foreign body or adhering to the atrial wall. From these morphologic points of view, the present case was considered to be a right atrial thrombus developing in situ.

From our literature survey, only a small percentage of patients who were studied by echocardiography showed right atrial thromboemboli. There are a few reasons why right atrial thromboemboli are not frequently detected. Most thromboemboli originate from systemic veins and pass rapidly through the right atrium and lodge in the pulmonary arteries. Another factor is imaging problems of intracavitary thrombus; that is, there is no good “echo window” to observe the whole right atrial and ventricular cavities, especially the right atrial free wall in which thrombus in situ frequently develops.

In this case, the stagnant and disturbed blood flow due to congestive heart failure was considered to have brought about activation of blood coagulation. This was suggested by the “smoke-like” echo noted around the thrombus. Moreover, the scoliosis and the pectus carinatum which caused diverticular right atrial dilatation were considered to be precipitating factors for thrombus formation. Thus, it is competent to suggest that the thrombus was developed during a recent congestive episode.

Concerning the therapy of a right atrial thrombus, the surgical removal is the first choice. We generally administer warfarin to prevent further development of thrombus. It might induce relative augmentation of effect of endogenous plasmin, so that the fresh thrombus was dissolved. Anticoagulant therapy for an immobile and fresh thrombus in situ, in contrast to metastatic and organized thromboemboli from peripheral veins, is a serious management problem in inoperative cases or in patients who reject operative therapy.

**REFERENCES**

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Long-Standing Indolent Blastomycosis at Internal Opening of Tracheostomy*

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An unusual case of long-standing endotracheal blastomycosis caused recurrent airway obstruction at a tracheostomy site.

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FITC = fluorescein isothiocyanate

Blastomyces dermatitidis is a dimorphic fungus which grows as a mycelium in moist, organic-rich soil and produces conidial spores. Inhalation of these spores may cause blastomycosis, one of the important endemic mycoses which affects humans, dogs, and horses in North America.1,2

In *vivo*, the fungus converts to its yeast form which may be identified in body secretions such as sputum or in body tissues. Blastomycosis is most common in the Ohio and Mississippi River valleys and in areas around the Great Lakes.

CASE REPORT

An 18-year-old man, previously in good health, went on a three-day canoe trip in late July 1984, along the Manistee River in Northern Michigan. He slept outdoors on the river bank. Three weeks later, he sustained severe head injuries and a fractured femur in a road accident in metropolitan Detroit, was rendered comatose, and intubated in the emergency room. A tracheostomy was performed nine days later following a craniotomy for debridement of brain tissue. He remained in a deep coma and required a mechanical respirator for several weeks following the accident. Commencing three months after the accident and during the following 2½ years, the patient required repeated removal of obstructive endotracheal granulations at the tracheostomy site which initially completely occluded the airway, preventing him from breathing or talking with the tracheostomy tube plugged. First visualized with a fiberoptic bronchoscope, the granulations were seen to arise anteriorly,

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FIGURE 1 (Upper). Section of endotracheal granulation shows acute and chronic inflammation with intraepithelial microabscesses (M) and Langhans' giant cells. There is squamous metaplasia and marked pseudoepitheliomatous hyperplasia (hematoxylin-eosin; original magnification × 160). (Lower left). High power of microabscess shows Langhans' giant cell containing two yeast forms marked with asterisks (hematoxylin-eosin, original magnification × 1600). (Lower right). Gomori's silver methenamine stain more clearly defines the yeast forms. Areas of broad-based budding, a characteristic feature of this fungus, are marked with arrows (original magnification × 1,600).

immediately above the stoma, measuring up to 3 cm diameter. They were removed using direct laryngoscopy with microscope and "suspension" with stripping. The rest of the trachea, larynx, and bronchi appeared normal, as did chest roentgenograms at the time of his emergency admission and on later occasions. Microscopic examination and cultures of endotracheal granulations removed in late 1984 proved negative for organisms, but a few yeast-forms were observed in tissue sections of granulations removed in February 1985 and were confirmed to be Blastomyces dermatitidis by the Centers for Disease Control, Atlanta, using FITC-conjugated antiglobulins specific for the tissue form of this fungus. Later biopsy specimens yielded positive fungal cultures and typical histologic features (Fig 1 and 2). Since late 1986, the patient has remained free of any clinical evidence of blastomycosis, despite his refusal to be treated with an antifungal agent such as ketoconazole or amphotericin B. However, smears of biopsy tissue from the inner margin of the apparently healed, but still patent, tracheostomy taken on April 18, 1988, revealed broad-budding yeast forms, (similar to those seen in Fig 2) consistent with blastomycosis.

DISCUSSION

Blastomycosis is most frequently a pulmonary disease which, in humans, is usually self-limiting, but which may become chronic and exhibit a high fatality rate and systemic...