Cardiac tamponade secondary to giant lymph node hyperplasia (Castleman's disease)

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Chest 1994;105;637-639

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platypnea is a result of postural changes causing an increased right-to-left shunt, most often through a patent foramen ovale. Another theory involves loss of the normal hypoxic vasoconstrictive response, causing increased shunting. Adolph et al described a patient with platypnea who had echocardiographic evidence of an intracardiac shunt with no pressure gradient in the setting of a pericardial effusion. They felt that the effusion distorted the architecture of the right atrium when the patient was standing in such a way as to direct the flow of blood returning from the inferior vena cava directly toward a patent foramen ovale.

To our knowledge, ours is the only reported case of platypnea in association with constrictive pericarditis. Although the patient had undergone a previous pneumonectomy, his symptoms could not be explained by this alone because they completely resolved after pericardiectomy. His diminished pulmonary reserve as a result of the prior pneumonectomy may have somehow rendered him more susceptible to the hemodynamic changes of constriction. This may be why platypnea is not seen in patients with constrictive pericarditis alone. Our patient had no evidence of an intracardiac shunt by supine and upright contrast echocardiography or by cardiac catheterization. We were unable to demonstrate a patent foramen ovale.

The reason for platypnea in our patient is not clear. One possible explanation is that with constrictive pericarditis he was very dependent on a high filling pressure to maintain an adequate cardiac output. With the patient in the upright position the right ventricular preload diminished, resulting in decreased right-sided cardiac output. This would cause diminished pulmonary arteriolar pressures and a greater proportion of zone I hemodynamics. As a result, there would be an increase in V/Q mismatch, increased work of breathing, and dyspnea. This cascade would be reversed in the recumbent position, with a return to higher filling pressures in the right side of the heart. After pericardiectomy, the cardiac output was less dependent on high preload, and a decrease in filling pressure on standing did not significantly alter his cardiac output. Therefore, pulmonary artery pressure was maintained, and there was no increase in zone I physiology.

There is much to be learned about this rare and complicated symptom. This report may add insight into possible pathophysiologic mechanisms of this phenomenon.

REFERENCES

Cardiac Tamponade Secondary to Giant Lymph Node Hyperplasia (Castleman's Disease)*

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A unique association of giant lymph node hyperplasia (Castleman's disease) and cardiac tamponade is presented. Although pleural effusions have been previously described with Castleman's disease, the authors believe this to be the first report of pericardial effusion and tamponade with this entity. The development of effusions may be due to an inflammatory syndrome sometimes seen with the plasma cell variant of this disease.

Castleman's disease is a rare entity characterized by benign hyperplasia of lymph nodes, occurring most commonly in the mediastinum. Clinically, it usually presents as an asymptomatic mass on routine chest radiograph. Although this condition has been associated with recurrent pleural effusions, symptomatic pericardial effusion has not, to our knowledge, been described to date with this disease.

CASE REPORT

A 54-year-old woman was admitted to an outlying hospital for progressive dyspnea over several days. Bilateral cervical and axillary lymphadenopathy had been present for 1 month, and painful erythema of the right breast had developed 1 week prior to admission. Within 12 h of presentation, the patient became hypotensive, and a computed tomographic scan of the chest showed a large pericardial effusion, as well as an anterior mediastinal mass (Fig 1). Pericardiocentesis yielded 250 ml of bloody fluid with immediate resolution of both the symp-

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CHEST / 105 / 2 / FEBRUARY, 1994 637

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Cardiac Tamponade and Giant Lymph Node Hyperplasia

The patient was transferred to our institution for further evaluation and care. Upon arrival, she was noted to have jugular venous distention and muffled heart sounds. In addition, there was bilateral axillary, supraclavicular, and cervical lymphadenopathy. The right breast was moderately tender and erythematous, but there was no palpable mass. A subsequent mammogram was negative for any mass. Laboratory values were notable for mild anemia and an elevated alkaline phosphatase concentration. An echocardiogram revealed a large pericardial effusion with diastolic collapse of the right ventricle, as well as multiple bands and septa suggestive of fibrinous adhesions. Repeat pericardiocentesis yielded 550 ml of bloody fluid.

Pericardectomy performed through a left anterior thoracotomy revealed hemorrhagic pericarditis with a thickened, inflamed pericardium that was adherent to the epicardium in several areas and thick fibrous bands and septa, indicative of an early constrictive process. There was a large, hard, yellowish anterior mediastinal mass overlying the ascending aorta. Part of this mass was removed in continuity with the pericardium. The anterior pericardium was resected to the phrenic nerve on each side, from the diaphragm inferiorly to the aortic reflexion superiorly. Cytology and cultures of the pericardial fluid were negative, and histologic study of the pericardium suggested nonspecific fibrinous pericarditis. The histologic diagnosis of the resected mediastinal mass was thymic tissue with acute and chronic inflammation and hyaline deposition. A right axillary lymph node biopsy specimen obtained several days later showed sheets of plasma cells widely separating atrophic germinal centers (Fig 2). Immunoperoxidase stain suggested a polyclonal, rather than a monoclonal, plasma cell population. This pattern was consistent with a mixed histologic form of Castleman’s disease as well. Retrospective review of the thymic tissue showed changes consistent with the diagnosis of Castleman’s disease. The patient was discharged with no specific therapy for the lymph node hyperplasia and is well 18 months later.

**DISCUSSION**

Castleman first described the entity of localized mediastinal lymph node hyperplasia resembling thymoma in 1956,1 and later characterized the disease further in a review of 81 cases.2 The main lesion was located in the mediastinum in 70 of these patients; however, extrathoracic sites of origin were noted in 8 cases. Histologically, the disease tends to occur in two distinct forms. The hyaline-vascular type accounts for approximately 90 percent of cases and is characterized by atrophic germinal centers separated by large areas of extensive capillary proliferation and fibrosis. The plasma cell type, which represents the remaining 10 percent of cases is identified by sheets of mature plasma cells in the interfollicular tissue and the presence of normal to large follicle centers. While the hyaline-vascular type tends to be diagnosed in asymptomatic individuals by routine chest roentgenogram, the plasma cell type is often associated with a clinical syndrome that consists of fever, fatigue, and peripheral lymphadenopathy. Laboratory abnormalities seen with this syndrome may include mild anemia, elevated sedimentation rate, hyperglobulinemia, hypoalbuminemia, and elevated alkaline phosphatase concentration. Because of its association with these clinical markers of an acute inflammatory response, it has been suggested that the plasma cell type represents an earlier, more active stage of the disease process, while the hyaline-vascular type represents a later stage.3 The underlying etiology of the disease remains speculative.

As for the case described herein, the histologic findings were mixed, but the clinical presentation and associated laboratory findings were certainly more consistent with the plasma cell variant of Castleman’s disease. There are several unique aspects of this particular case. First is the presence of massive pericardial effusion and cardiac tamponade. There have been previous reports of pleural effusion in association with Castleman’s disease,4,5 but none to date has documented an association with pericardial effusion. Acute pericarditis, which was documented histologically, can apparently occur as one manifestation of the generalized inflammatory syndrome related to plasma cell histology. The effusion results from an inflammatory exudate as well as impaired reabsorption by the
inflamed serous membrane. Second, no specific treatment was rendered for the mediastinal mass. The accepted standard of therapy in Castleman's disease is surgical excision, which is usually performed as both a diagnostic and a therapeutic maneuver and is uniformly curative. Although there have been rare reports linking Castleman's disease with Kaposi's sarcoma, the disease is not considered premalignant, nor have there been any reported recurrences after surgical resection. Radiation and chemotherapy have generally not been successful when applied to patients who could not undergo surgical treatment.

The diagnosis in this case was made by axillary lymph node biopsy because of an erroneous initial interpretation of the excised thymic tissue as normal. Since the patient had no symptoms related to local compression or infiltration, repeat mediastinotomy was not recommended. To this date, the patient has remained well.

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