

Original article

Constrictive Pericarditis: Etiologic Spectrum, Patterns of Clinical Presentation, Prognostic Factors, and Long-term Follow-up

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ABSTRACT

Introduction and objectives: Some reports have described a change in the etiologic spectrum of constrictive pericarditis. In addition, data on the relationship between its clinical presentation and etiology are lacking. We sought to describe the etiologies of the disease, their relationship with its clinical presentation and surgical findings, and to identify predictors of poor outcome.**Methods:** We analyzed 140 consecutive patients who underwent surgery for constrictive pericarditis over a 34-year period in a single center.**Results:** The etiology was idiopathic in 76 patients (54%), acute idiopathic pericarditis in 24 patients (17%), tuberculous pericarditis in 15 patients (11%), purulent pericarditis in 10 patients (7%), and cardiac surgery, radiation and uremia in 5, 3 and 2 patients respectively (4%, 2% and 1%). Mean duration of symptoms before pericardiectomy was 19 months (standard deviation, 44 months), the most acute presentation being for purulent pericarditis (26 days [range, 7-60 days]) and the most chronic for idiopathic cases (29 months [range, 4 days-360 months]). Perioperative mortality was 11%. There was no difference in mortality between etiologies. Median follow-up was 12 years (range, 0.1-33.0 years) in which 50 patients died. In a Cox-regression analysis, age at surgery, advanced New York Heart Association functional class (III to IV) and previous acute idiopathic pericarditis were associated with increased mortality during follow-up.**Conclusions:** Most cases of constrictive pericarditis are idiopathic. Cardiac surgery and radiation accounted for a minority of cases. Etiologic investigations are warranted only in acute or subacute presentations. Age, advanced functional class, and previous acute idiopathic pericarditis are associated with increased mortality.

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Pericarditis constrictiva: espectro etiológico, presentaciones clínicas, factores pronósticos y seguimiento a largo plazo

RESUMEN

Introducción y objetivos: Algunos estudios han descrito un cambio en el espectro etiológico de la pericarditis constrictiva. Además, no hay datos sobre la relación entre la forma de presentación clínica y la etiología. El objetivo de este estudio es describir las etiologías de la enfermedad, su relación con la forma de presentación clínica y los hallazgos quirúrgicos, así como identificar los factores predictivos de una mala evolución.**Métodos:** Se analizó a un total de 140 pacientes consecutivos a los que se practicaron intervenciones quirúrgicas por pericarditis constrictiva en un mismo centro en un periodo de 34 años.**Resultados:** La etiología fue idiopática en 76 pacientes (54%) y tras pericarditis aguda idiopática en 24 (17%), pericarditis tuberculosa en 15 (11%), pericarditis purulenta en 10 (7%) y cirugía cardíaca en 5 (4%), radioterapia en 3 (2%) y uremia en 2 (1%). La duración media de los síntomas antes de la pericardiectomía fue de 19 meses (desviación estándar: 44 meses); la forma de presentación clínica más aguda fue la de las pericarditis purulentas (26 [intervalo, 7-60] días) y la más crónica, la de los casos idiopáticos (29 meses [4 días-360 meses]). La mortalidad perioperatoria fue del 11%. No hubo diferencias en la mortalidad según etiologías. La mediana de seguimiento fue de 12 (0,1-33,0) años, durante los cuales fallecieron 50 pacientes. En un análisis de regresión de Cox, la edad en el momento de la operación, la clase funcional

Palabras clave:

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Pericarditis constrictiva

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de la *New York Heart Association* avanzada (III–IV) y los antecedentes de pericarditis aguda idiopática se asociaron a una mayor mortalidad durante el seguimiento.

Conclusiones: La mayoría de los casos de pericarditis constrictiva son idiopáticas. La cirugía cardíaca y la radioterapia causan una minoría de los casos. Las presentaciones aguda y subaguda merecen un estudio etiológico. La edad, la clase funcional avanzada y la pericarditis aguda idiopática previa se asocian a mayor mortalidad.

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Abbreviations

CP: constrictive pericarditis
CS: cardiac surgery
CT: computed tomography
PE: pericardial effusion

INTRODUCTION

Since constrictive pericarditis (CP) was first described in 1669 by Lower,¹ some clinical aspects may have changed. Some studies² have recently suggested that the etiologic spectrum has changed in the last decades, basically due to an increase in the number of cases secondary to cardiac surgery (CS) or previous radiotherapy. However, this finding could be misleading. First, these studies were conducted in centers with a high cardiac surgical activity,³ which could introduce some bias in the etiologic spectrum. In addition, the relationship between symptom duration before pericardiectomy and the etiology of CP was not described in detail. Finally, previous studies usually did not include accurate information on the different diagnostic procedures or imaging studies used to diagnose CP.

Our center, a referral center for pericardial diseases, has used a standardized protocol for diagnosis and management of pericardial disease^{4,5} since 1975. In the present report we describe the etiologic spectrum, the relationship between the chronicity of clinical symptoms and the etiology of CP, the use of the different diagnostic procedures, and predictors of poor outcome, in a series of 140 consecutive patients with CP who underwent surgery in our institution in the last 34 years.

METHODS

We conducted a retrospective study in a single tertiary center. The study was approved by the ethics committee of our institution. A total of 140 patients underwent pericardiectomy in our institution (*Hospital Universitari Vall d'Hebron, Barcelona, Spain*) for confirmed CP between January 1978 and May 2012. The clinical records were individually reviewed by a single investigator (Andreu Porta-Sánchez), devoting special attention to the presence of inflammatory systemic diseases, previous episodes of acute pericardial diseases, thoracic or mediastinal irradiation or previous CS, and the clinical and imaging studies performed. The time elapsed from the potential causative illness to the onset of symptoms of CP was quantified as well as the time to pericardiectomy. Surgical features were also recorded, with special attention to the involvement of the parietal and visceral layers. Mortality and follow-up data were obtained through outpatient clinic visits or by the electronic clinical records or by phone. Data from the Spanish National Institute of Statistics⁶ were

obtained to generate an age and sex-matched population, and an estimated Kaplan Meier survival curve was generated to compare the long-term overall mortality in our patients with the general population.

Definitions

Idiopathic CP was diagnosed when no apparent etiology was identified. Postacute idiopathic pericarditis was defined as CP occurring after a well-defined episode of acute idiopathic pericarditis (typical chest pain, pericardial friction rub, serial changes in the ST segment of the electrocardiogram). Tuberculous CP was defined when Koch bacillus was isolated from a pericardial specimen or when caseous granulomas were identified in a biopsy specimen. Purulent CP was diagnosed when pericardial fluid was macroscopically purulent or had a high polymorphonuclear content (> 90%). Postsurgical CP was diagnosed when CP was developed after an open-heart surgery. Radiotherapy CP was defined by the development of CP after a therapeutic chest radiation and after the exclusion of other causes of CP. In an arbitrary manner, the terms acute, subacute and chronic CP were applied, respectively, to patients who required pericardiectomy before 3 months, between 3 and 6 months, or after 6 months of the onset of clinical symptoms. Pericardial calcification was defined as the presence of calcification on a chest X-ray. Left ventricular systolic function was considered abnormal if left ventricular ejection fraction was estimated below 50%. The following echo-Doppler findings were considered diagnostic: the presence of pericardial thickening and notch of the interventricular septum, changes in E velocity superior to 25% related to respiratory movements, a rapid E wave deceleration, suprahepatic vein flow with “W” morphology or predominance of the diastolic component with diastolic inversion during expiration. The amount of pericardial effusion (PE) was classified as mild (< 10 mm of echo-free space in anterior and posterior pericardial spaces), moderate (10–20 mm) or severe (> 20 mm). Additional imaging studies were performed to confirm pericardial thickening or calcification (computed tomography [CT]) or to exclude an infiltrative cardiomyopathy (magnetic resonance imaging). If diagnosis of CP was still unclear after the imaging studies, a right and left heart catheterization was performed. Pericardiectomy was performed through median sternotomy in all cases. Although there have been several surgical advances in the last 30 years, the surgical approach for the treatment of CP has not substantially changed over this period. A wide pericardiectomy was attempted (phrenic-to-phrenic and diaphragmatic pericardium) and was successfully achieved in 95% of cases. Perioperative mortality was defined as death occurring during surgery or within the first 30 days of hospitalization.

Statistical Analysis

The 1-way analysis of variance test was used for comparisons of continuous variables between etiologic groups. The chi-square

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