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Review article

Current concepts of diagnosis and management of pericardial cysts

Sandeep Kumar Kar*, Tanmoy Ganguly

Department of Cardiac Anesthesiology, Institute of Post Graduate Medical Education and Research, Kolkata, India

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ABSTRACT

Pericardial cysts are rare with an incidence of about 1 in every 100,000 persons and one in 10 pericardial cysts may actually be a pericardial diverticulum. Pericardial cysts and diverticula share similar developmental origin and may appear as an incidental finding in chest roentgenogram in an asymptomatic patient. CT scan is considered as best modality for diagnosis and delineation of the surrounding anatomy. Cardiac MRI is recommended in the evaluation of the compressive effects caused by the pericardial cysts. The authors recommend echocardiography for serial follow up and image guided aspiration of the pericardial cyst in presence of compressive effects leading to cardiovascular and airway symptoms. A systematic approach is desirable for management of pericardial cysts depending on size, shape and compression effects, symptoms and easy access to serial Echocardiographic follow up. However, pericardial diverticulum may not be differentiated from cysts by the above testing, and only identified at surgery.

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Contents

1. Introduction

Cystic lesions within the pericardial space are a rare entity and comprise 7% of the mediastinal masses and 33% of mediastinal cysts.[1,2] The incidence of a pericardial cyst is 1 in 100,000 populations and most of the pericardial cysts presenting as mediastinal opacity are detected incidentally.[3–6] They are usually found in the third or the fourth decade of the life and male and female are affected equally.[5] In 70% of the cases, these cysts are located in right cardiophrenic angle, in 22% cases in the left cardiophrenic angle and in 8% cases are located in the posterior or the anterior-superior part of the mediastinum.[7] Most of the cases (50–75%) are asymptomatic and are diagnosed incidentally during radiological investigations ordered as routine investigation for other causes of illness.[6–11] Symptoms may appear due to compression of the nearby structures, such as heart, great vessels, oesophagus and the tracheobronchial tree.[11] Pericardial cysts are described in the medical literature under various terminologies like: le kyste pleuropericardique (Jeaubert de Beaujeu et al., 1945; Roche et al., 1954), pleural cyst, pericardial cyst, pericardial

* Corresponding author.
E-mail address: sndpkar@yahoo.co.in (S.K. Kar).

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coelomic cyst (Lambert et al., 1940), springwater cyst (Greenfield et al., 1943), mesothelial cyst (Churchill and Mallory, 1937), and thin-walled cyst.[12]

2. History

Preliminary reports of pericardial cysts date back to middle of 19th century when the pathologists encountered the initial cases on post mortem examination.[13] Advances in radiographic imaging made antemortem diagnosis possible and Le Roux et al. reported three cases in 300,000 people in a mass chest roentgenography campaign in Edinburgh in 1958.[14] Progress in the field of thoracic surgery ushered a new era in management of these lesions and Otto Pickhardt et al. from Lenox Hill Hospital performed the first resection of pericardial cyst in New York in 1931[15] in a 53 year old woman. The first reported incidence of pericardial diverticulum was presented by T. Hart of the Park Street School of Medicine in Dublin, in 1837.[16] Subsequently a new concept of origin of pericardial cysts and diverticula was proposed, according to which pericardial cysts and diverticula represent different stages of a lesion with a common embryonic origin. Greenfield et al. coined the term 'springwater cysts' because of the presence of thin, transparent cyst wall and crystal clear fluid content within the pericardial cyst.[17] Surgical approach to the pericardial cysts and diverticula has undergone several refinements with the present evidence based medical diagnosis and treatment approach. Currently video assisted thoracoscopic surgery is considered as the most promising technique in the diagnosis and in the management of these lesions. A brief chronology of events in discovery and gradual evolution of different modalities of management is outlined in Table 1.

3. Pericardial cyst and diverticula

Although radiologically a lesion may appear to be a pericardial cyst, in 10% of the time, it is actually a diverticulum.[18] Although dissimilar on anatomical basis, both pericardial cyst and diverticula are considered as a sequelae of common embryogenesis going wrong and are usually discussed together by most embryologists.[18–24.] Pericardial cysts and diverticula usually arise due to herniation through a structural defect in the pericardium.[19,20] Rohn et al. first described similar origin of both the lesions and concluded that the pericardial cyst probably is a remnant of a diverticulum whose communication to the pericardial cavity has been obliterated.[23] To assure that it is truly a "cyst", it might be necessary to trace the try to identify communicating channels during dissection of the pericardial cysts and if found be ligated to prevent a diverticulum from reoccurring.[18] Pericardial diverticula may be congenital or acquired. Congenital cases may result from a failure in the fusion of one of the mesenchymal lacunae that normally combine to form the pericardial sac. Acquired pericardial diverticula may be due to sequelae of pericardial diseases and effusion.[18–22]

4. Origin

Pericardial cysts are usually congenital in origin but other causes of origin of pericardial cysts have also been described in literature (Table 2). Pericardial cysts usually arise from failure of fusion of one of the mesenchymal lacunae that form the pericardial sac.[5] Adrian Lambert suggested that the cyst and diverticulum embryologically originate from the disconnected mesenchymal lacunae which normally unite to form the pericardial coelom.[27] Mazer described the fluid shift from pericardial diverticula to pericardial sac and attributed this as the cause of congestive chest symptoms and chest discomfort.[28] Lillie et al. expounded the origin of the pericardial cysts by the concept of differential persistence and graded constriction of ventral recess of the pericardial coelom. Persistence of the ventral recess of the pericardial coelom forms the diverticulum, constriction of the proximal part of the persistent recess accounts for either a diverticulum with a narrow neck or results in the origin of a pericardial cyst in communication with the pericardial cavity and complete closure of the proximal recess forms the pericardial cyst.[29] Prenatal diagnosis of pericardial cyst is made possible with ultrasound examination beyond 14th week of gestation.[30] Cases of spontaneous regression of pericardial cysts have also been described in literature.[31] Inflammatory cysts and pseudocysts appear due to loculated pericardial effusion.[6] Isolated hydatid cyst of pericardium is extremely rare and they are usually found in association with myocardial cysts or hydatid cysts in the liver and the lungs.[32,33]

5. Clinical presentation

Patients with pericardial cysts are mostly asymptomatic (50–75% cases) [5–11](50–75%cases) [5–11] and the diagnosis is usually an incidental finding in chest X-ray. Symptoms usually appear when the cyst compresses on a nearby structure, or undergoes complications[39–42] (vide Table 5) . Common symptoms include chronic cough, chest pain, dyspnea and a feeling of retrosternal pressure.[40,41] Abdul- Mannan Masood et al. described a case of a large pericardial cyst (11 cm × 11 cm) in a patient presenting with right shoulder discomfort radiating to the left shoulder, with associated heaviness in the substernal area along with shortness of breath.[43] Recurrent attacks of palpitation due to cardiac dysrrhythmias and frequent lower respiratory tract infections have been described in literature.[5] Unusual presentations of pericardial cysts include recurrent syncope,[44] pneumonia,[45] congestive heart failure and sudden cardiac death.

Patients with pericardial diverticula may have atypical symptoms that cannot be explained. There is typically no evidence of

Table 1
A brief chronology of events in discovery and gradual evolution of different modalities of management.

Year	Events
1837	T. Hart of the Park Street School of Medicine in Dublin described the first case of a pericardial diverticulum on autopsy[16]
1903	Rohn, from the Charles University of Prague first published a case series comprising of four diverticula and one cyst based on autopsy finding of these lesions. The interrelationship between pericardial diverticulum and cyst sharing a common embryonic origin was first recognised in this case series.[23]
1931	Wallace Yater (Georgetown University) detailed the radiological appearance and the differential diagnosis of pericardial cysts[25]
1931	Otto Pickhardt, at Lenox Hill Hospital in New York performed first surgical removal of pericardial cyst[15]
1937	First pneumogram of pericardial cyst was performed by E. H. Cushing[26]
1940	Adrian Lambert first suggested similar embryological origin of pericardial cyst and diverticula from disconnected mesenchymal lacunae, which later unite to form the pericardial coelom[27]
1943	First resection of a pericardial diverticulum by Richard Sweet at Massachusetts General Hospital in Boston[29]
1943	Greenfield et al introduced the term 'Springwater cyst'[17]
1958	Le Roux reported the incidence of three cases of pericardial cysts in 300,000 people in a mass X-ray campaign in Edinburgh[14]

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