Chronic Primary Chylopericardium

Report of a Case and Review of the Literature

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SUMMARY

The case of a 36-year-old white man who had recurrent milky pericardial effusion and repeated pericardiocenteses over a period of 1 year with minimal disability or symptoms is presented. Surgical management included thoracotomy with ligation of all structures resembling lymphatic channels in the chest in several places and hemi-pericardiectomy. Neither the source of chyle in the pericardium nor the etiological agent of this condition could be found. Criteria for successful management of this condition are re-emphasized.

ADDITIONAL INDEXING WORDS:
Pericardiectomy Lymphatics Pericardiocentesis Pericardial effusion

The isolated accumulation of chyle in the pericardial space has been recognized as a clinical entity only since 1954. At that time a mediastinal hygroma (lymph-angiomatous hamartoma) in association with chylopericardium in a 31-year-old woman was reported by Groves and Effler. Although no direct communication between the tumor and either the pericardium or the thoracic duct could be identified at surgery, it was assumed that such communication did exist. Since that time two additional cases of similar tumor and isolated chylopericardium have been reported, as well as four cases of chylopericardium in which no such possible etiological mechanism could be found.

The term “primary chylopericardium” was used by Groves and Effler in the first paper and it seems proper to retain this term until more exact etiological mechanisms can be identified.

The value of single case reports in leading to better management of patients with rare clinical entities is well shown in this condition.

The seven previously reported cases of chylopericardium without chylothorax were all extensively investigated and the correct surgical approach has been fairly well worked out. There is no reason to believe that such cases did not exist in the past. However, the condition is not discussed in standard textbooks prior to 1954, even though Christian’s 1947 edition of Osler’s Principles and Practice of Medicine contains the single sentence, “In rare cases the serum (i.e., pericardial fluid) has a milky character, chylopericardium.”

This same sentence appeared in Osler’s first edition in 1892, with no elaboration or references. No other mention of this condition could be found during a search through several other medical and pathological textbooks of the nineteenth and early twentieth centuries.

This report deals with the eighth such case in the English literature. Similar to the previously reported cases, this patient presented a diagnostic problem for many months. Although our follow-up has been brief, surgery has apparently resulted in complete cure. Neither tumor nor abnormal lymphatic communications could be identified at surgery. This case differs somewhat from the other seven in two main respects: (1) the relatively asymptomatic state of the patient, and (2)
the massive amounts of chyle obtained over a period of 13 months. Although the recently reported case by Hudspeth and Miller\(^7\) suggests a possible duration of 6 years without symptoms; severe symptoms developed in their case after the first pericardiocentesis.

Report of Case

W. R., a 36-year-old white man, a machinery salesman, was referred to the Cardiovascular Division of the Department of Medicine, University of Alabama Medical Center, for recurrent pericardial effusion and was admitted October 25, 1965. The patient had been in good health until December 1963, when he noted the onset of epigastric and substernal fullness and discomfort, "as if I had eaten a huge meal," which was not related to meals, time of day, or physical effort. He attributed this symptom to a possible "ulcer" and treated himself intermittently with various antacids and other preparations without benefit. The sensation of fullness became persistent during the following 10 months with only minimal increase in severity. No other symptoms were noted and certainly none suggestive of possible cardiovascular pathology.

He finally consulted his physician in October 1964, for "stomach trouble." A massive pericardial effusion was discovered, the patient was hospitalized, and pericardiocentesis produced 700 ml of fluid which the patient described as being "like vanilla milk shake." The discomfort was promptly relieved with the tap. Within a few days the symptom recurred and reaccumulation of fluid was noted. A repeat tap 2 weeks later produced another 700 ml of similar fluid. Routine cultures and cultures for acid-fast bacilli and fungi of the fluid were negative, and the patient was empirically treated with antituberculous drugs for 4 months, followed by 2 months of steroid therapy without apparent diminution of the rate of fluid reaccumulation. From then until his present admission, the patient was treated symptomatically with repeated taps. Whenever his symptoms recurred and advanced to the stage indicating reaccumulation of a large quantity of fluid, he was admitted to the local hospital, tapped, and discharged the next morning asymptomatic, to return to work. During this entire period, except for his first hospitalization, although he had a total of seven pericardiocenteses yielding as much as 2,900 ml on one occasion, he continued his full-time employment without interruption and continued his normal everyday activities.

On admission to the University Hospital, the patient was essentially asymptomatic except for his usual symptoms of epigastric and substernal fullness and discomfort. His most recent tap was approximately 1 month prior to this admission. He denied fever, chills, joint pains or swelling, weakness, headaches, easy fatigability, exertional dyspnea, orthopnea, fluctuations of weight, peripheral edema, night sweats, or any other symptoms.

The past medical history was remarkable only in that the patient was involved in an automobile accident in January 1963, approximately 11 months before the onset of symptoms. At that time he suffered only a minor whiplash-like injury to the neck resulting in mild stiffness of his neck for a couple of weeks followed by complete recovery. He did not feel then that there were any indications for medical evaluation and none was sought. He denied any memory of blows or other trauma to the chest or any pain or tenderness of the chest or chest wall.

Physical examination revealed a well-developed, healthy-appearing adult male lying flat in bed in no apparent discomfort. Oral temperature was 99°F, pulse was 94 per minute and regular, respirations were 14 per minute, and the blood pressure was 120/95 mm Hg in both arms with no demonstrable paradoxical pulse. The veins of the neck were full but not significantly distended above 30.\(^6\) The Kussmaul sign (inspiratory expansion of the cervical veins) was not elicited. The thyroid was not palpable. The lungs were clear. An apical impulse was not palpable. The left border of the heart was percussed at the left midaxillary line and the right border, at the right midclavicular line. The heart sounds were audible but distant and no murmurs, pericardial friction rubs, or gallops were appreciated.

Figure 1

Chest roentgenogram taken on admission, October 25, 1965.
The peripheral pulses were adequate and equal bilaterally. The liver and spleen were not palpable, and there was no evidence of ascites or peripheral edema.

Fluoroscopic studies revealed a markedly enlarged cardiac shadow with the configuration of a massive pericardial effusion (fig. 1). The electrocardiogram was within normal limits. The white cell count was 8,900 with normal differential. The corrected erythrocyte sedimentation rate was 7 mm per hour and packed cell volume was 50%. Other laboratory findings were transaminase (SGOT) 15 units, 2-hour postprandial blood sugar 80 mg%, serum calcium 4.8 mEq/L, serum phosphorus 3.2 mg%, total serum cholesterol 233 mg%, blood urea nitrogen 12 mg%, serum protein 7.7 g% with normal electrophoretic pattern, and ASO titer 125 Todd units. L. E. cell preparations were negative on three occasions, and skin tests for tuberculosis, histoplasmosis, and coccidioidomycosis were negative. The vital capacity was 2.4, 2.5, and 2.6 L on three occasions which was 60% of the predicted normal. The central venous pressure was 80 mm of saline and there was no change with the Valsalva maneuver to suggest cardiac tamponade. Uralysis was within normal limits.

On October 26, 1965, a transthoracic pericardiocentesis under local anesthesia was performed without difficulty and 1,100 ml of milky fluid was obtained. No attempt was made to remove all fluid possible. Two hundred milliliters of air was introduced into the pericardial sac and fluoroscopy then revealed a thin pericardium with a large pericardial effusion being still present (fig. 2). The fluid had a specific gravity of 1.036 and a total protein of 5.9 g% with an electrophoretic pattern identical with that of the serum protein. There were 8,000 fresh red blood cells and 280 lymphocytes per cubic millimeter. Values for amylase were 186 U, for cholesterol 104 mg% and for triglycerides 1,640 mg%. When equal volumes of diethyl ether and the fluid were mixed in a tube, almost complete clearing of the milkiness of the fluid occurred. Cultures for acid-fast bacilli, fungi, and bacteria were negative. Histological studies on cell block preparations revealed no tumor cells.

The patient remained afebrile during hospitalization and had an essentially benign course. The diagnosis of primary chylopericardium was made and surgery was recommended. Because of pressing personal business, the patient was discharged on October 29, 1965, to return for readmission and surgery as soon as feasible.

He was readmitted to the Surgical Service on November 8, 1965, and 2 days later underwent exploratory thoracotomy by Dr. Merrill Bradley. Upon opening the chest, the pericardial sac was seen to be filled with a considerable amount of milky fluid (fig. 3). It was opened and evacuated of 2 to 3 L of milky, odorless fluid. The pericardium appeared otherwise normal without significant thickening or any areas of adhesion to the heart, and a hemipericardectomy was performed. No lymphatic communications with the pericardial space could be found nor could a definite thoracic duct structure be identified at the level of the pericardium even after several injections of methylene blue in the peri-diaphragmatic area. No lymphadenopathy, lymphangioma, or apparent malformations were noted. Structures which resembled lymphatic channels in the thorax and the site of entrance of the thoracic duct into the left subclavian vein were ligated in several places and at several levels in the thorax. The postoperative course was uneventful except for an area of atelectasis in the lower lobe of the left lung and partial paralysis of the left hemidiaphragm during the immediate postoperative period. No reaccumulation of fluid (or chyle) was noted 6 months after surgery and the patient was asymptomatic (fig. 4). Microscopic examination of the pericardium showed chronic nonspecific pericarditis with hyalinized nodular areas thought to be sites of previous pericardial taps.

Discussion

This case is almost identical to the cases reported by Knight and Hudspeth and Miller which also demonstrated the remarkable

Figure 2

Pneumopericardium. Roentgenogram taken October 26, 1965, immediately after aspiration of 1,100 ml of fluid and injection of 200 ml of air. Note air-fluid level within thin and enlarged pericardium.

Circulation, Volume XXXV, April 1967
lack of symptoms or disability to the patient in the presence of massive pericardial effusion. Although the other five patients whose cases have been reported also presented with relatively mild symptoms early in the course of the disease, they all progressed with time to the state of cardiac tamponade with disabling and serious manifestations.\textsuperscript{1-5} From the date of diagnosis to surgery, a period of 13 months, our patient had undergone eight pericardiocenteses in the following periods: October, November, and December 1964 and January, March, June, September, and October 1965. The last tap was mainly for diagnostic purposes upon his first admission to this hospital and not because of any significant symptoms. The only indication for the taps was the recurrence of the patient's symptom of epigastric and substernal fullness and discomfort. With each tap this discomfort was completely relieved, and the patient did not lose a single day of work during the year prior to his hospitalizations for definitive treatment. During the periods between pericardiocenteses, the patient enjoyed his normal everyday activities without interruption or significant discomfort. It is interesting to note

\begin{figure}[h]
\centering
\includegraphics[width=0.8\textwidth]{figure3.png}
\caption{During surgery, showing chyle-filled pericardium.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=0.8\textwidth]{figure4.png}
\caption{Chest roentgenogram made approximately 6 months after surgery.}
\end{figure}
from the approximate dates of these taps that the
time intervals between them were becoming
longer, yet, according to the patient the
amounts of fluid removed were somewhat
increased with each succeeding tap. This sug-
gested gradual reaccumulation of the fluid,
hence affording time for accommodation of
the pericardial sac and allowing it to hold
progressively larger volumes of fluid each
time before causing recurrence of the symp-
toms. This probably explains the paucity of
symptoms suggestive of tamponade in spite
of the presence of such massive amounts of
pericardial fluid.

In three of the reported cases1–3 lymph-
angiomatosus hamartomas were found, but no
causative pathology could be found in the
other four.4–7 In the case reported by Naef,4
there was a history of recent trauma and of
an inflammatory process but there were no
pathological findings on surgery to implicate
or suggest either as the cause.

In almost all cases, including this one, no
source of chyle in the pericardium could be
identified at surgery. The case reported by
Miller and associates,3 however, is an excep-
tion. In that case, the author described rapid
reaccumulation of chyle in the pericardium
at the rate of 50 ml in 10 minutes after the
pericardium was initially opened and the
fluid removed. The fluid appeared to flow
from many areas within the pericardium.
There have been two deaths2,3 and these
occurred in the patients who did not have
ligation of the thoracic duct. In both cases
frequent recurrent chylothorax and chyloperic-
ardi um complicated the entire postoperative
course appearing almost immediately after
surgery and continuing until death about 3
months later in two patients. The other six
had ligation of the thoracic duct or all major
lymphatic channels in the thorax or both, plus
a pericardial window or a partial pericardiec-
tomy. Our patient had ligation of all struc-
tures in the thorax which resembled lymphatic
channels in several places and also hemiperi-
cardectomy. No thoracic duct could be iden-
tified in our patient even after repeated
injections of lipophilic dyes in the peridia-
phragmatic region. The patient remains
asymptomatic 7 months after surgery, with
no evidence of reaccumulation of fluid.

We can only re-emphasize what the other
authors have already stated in regard to the
management of primary chylopericardium. To
assure success, the thoracic duct and all lym-
phatic channels in the thorax should be ligated
in several places, a pericardial window and preferably a partial pericardectomy should
be done to ensure adequate drainage and to
prevent the possible development of con-
strictive pericarditis, and a thorough search
for any causative factors should be conducted
and the elimination of such when encoun-
tered. Failure to comply with these require-
ments in the management of primary chylo-
pericardium will only invite disaster.

Acknowledgment

We wish to extend our appreciation to Dr. Tinsley
R. Harrison for his suggestions and comments in
reviewing this manuscript.

References
chylopericardium. New Eng J Med 250: 520,
1954.
2. Steatton, V. C., and Grant, R. N.: Cervico-
mediastinal cystic hygroma associated with
3. Miller, S. W., Pruett, H. J., and Long, A.: Fa-
tal chylopericardium caused by hamartomatous
lymphangiomatosis. Amer J Med 24: 951,
1959.
5. Madison, W., Jr., and Logue, B.: Isolated
(primary) chylopericardium due to anomalous
communications with the thoracic duct of
unknown causation. Amer J Med 22: 825,
1957.
(primary) chylopericardium: Diagnosis and
surgical treatment. J Thorac Cardiov Surg
51: 528, 1966.
8. Christian, H. D.: Principles and Practice of
Medicine (originally by Sir William Osler),
ed. 15. New York, D. Appleton, Century Co.,
1947, p. 1026.


**Famous Facetiae**

In the address, his Baltimore valedictory, delivered on Washington's birthday, February 22, 1905, Osler spends a good deal of time discussing the intellectual infantilism as well as the precocious senility or progeria which readily besets any teacher whose career is spent too much in one place. Today we are more likely to be corrupted by the Brownian motion of travel than by sessile inertia. Instead of quinquennial brain dusting, our trips and tours occur by month or week, by day or hour. This was not so during Osler's time. The urge to move, to accept new challenges, and to keep out of the deepening ruts of uniformity, was on Osler's mind. He had long held that the great and seminal work of the world was and could only be done by youth. In the address, he said, "The teacher's life should have three periods, study until 25, investigation until 40, profession until 60, at which age I would have him retired on a double allowance. Whether Anthony Trollope's suggestion of a college and chloroform should be carried out or not, I have come to be a little dubious as my own time is getting short."—William B. Bean: Osler, the Legend, the Man and the Influence. Canad Med Ass J 95: 1035, 1966.
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Circulation. 1967;35:777-782
doi: 10.1161/01.CIR.35.4.777

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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