Cardiospasm

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I. ABSTRACT

CARDIOSPASM

Clarence Dennis.

Definition:

Chevalier Jackson defines cardiospasm as "stenosis of the lower end of the esophagus which is primarily functional, but which is often complicated in the later stages by organic stenosis, and is always accompanied by more or less dilation of the esophagus if it is more than a few months duration."

Incidence

Cardiospasm rarely defined before x-ray came into general use.

Vinson reported 683 cases at Mayo Clinic from 1908 until 1929.

Freeman - 150 cases at Digestive Clinic of Johns Hopkins Hospital from 1912 to 1933.

At University of Minnesota Hospitals, there have been 7 cases in the past 30 months.

Pathology

Few pathological studies have been made, first because cardiospasm is rare and secondly because few patients die from the disease.

The gross picture is that of dilation of the lower two-thirds of the intrathoracic esophagus, with or without postmortem evidence of organic stenosis of the lower end of the esophagus. Usually, the walls of the cardia and the dilated portion of the esophagus are thickened. The longitudinal and circular muscles are hypertrophied. Some of the thickening may be inflammatory in origin. There is often warty, leathery thickening of the mucosa.

Microscopically, the commonest finding is degeneration of Auerbach's plexus, usually leaving only scarred remnants of former ganglion cells, with groups of round cells about them. The vessels are thin walled and dilated. The muscle may show hypertrophy, fatty degeneration, or hyaline changes. The esophagus above the dilation is usually normal.

Pathogenesis

The origin of the condition probably is primarily a disturbance in the peristalsis of the esophagus. Apparently the cardia fails to relax when the food boli reach it. Attention has been drawn to the anatomy of the cardiac esophagus (cardiac antrum). This is bounded above by a slight constriction formed by a sphincter elaborated from the intrinsic esophageal musculature. It is limited below by the inferior anatomical cardia, a ringlike fold of mucosa partially separating the lumen of the cardiac antrum from that of the stomach. The antrum is about 5 cm. long and is one-half above and one-half below the diaphragm. The muscles of the diaphragm form a third constriction, approximately in the middle of this cardiac antrum. The upper sphincter is supplied by the sympathetic, the phrenic sphincter by the phrenic nerve.

Most observers concede that the cardiac obstruction is primary and the hypertrophy and dilation above this is secondary.

Theories of genesis

1. Congenital
2. Malformation
3. Infection
4. Irritation of poorly chewed food
5. Local irritation
6. Nerve imbalance
7. Hypertrophy of diaphragm.

Comment

Many believe that cardiospasm is congenital and that the symptoms do not appear as long as there is compensation through muscular hypertrophy. Congenital twists and turns in the esophagus and abnormalities in the "hepatic esophageal tunnel" have also been suggested. Others contend that the origin lies in irritation or infection within the esophagus or cardia. Guisez blames the onset to the swallowing of poorly

chewed food, claiming that boli are held above the normal constriction of the cardia because of their large size; their arrest at this point then causes esophagitis and reflex spasm. He points to the increased incidence of cardiospasm in old patients, most of whom are edentulous. The theory of local irritation is also backed by the large incidence of cardiospasm with peptic ulcers, and the occasional case with gastric carcinoma, with diaphragmatic hernia, etc.

The role of the autonomic nervous system has been particularly stressed. The nervous control of the cardiac sphincter is derived from the vagus through Auerbach's plexus and from the sympathetics. Section of the sympathetic allows relaxation. Section of the vagus allows unopposed sympathetic action and results in increased tone. Rosenheim has even suggested that the whole condition is due to atony of the esophagus due to vagus injury such as may occur with lead poisoning. It is interesting that no such dilation of the esophagus occurs with an organic stenosis. Fulde has shown that the pressure in the cardiac region can be raised by irritation of the vagus, the sympathetic, or the phrenic nerve. Rieder cut the vagi in a case of cardiospasm and the symptoms became more severe. When he cut the vagi in dogs, he obtained a microscopical and gross picture identical to that in cardiospasm. He believes the vagal damage may be anywhere from the brain stem to the nerve endings. Working on the theory that sympathetic preponderance is the cause of cardiospasm, Craig did sympathetic ganglionectomies from 8_C to T_3 bilaterally with complete cure of an otherwise intractable case.

Many writers, notably Jackson, believe that the obstruction does not come from the sphincter but rather from the diaphragmatic muscle surrounding the cardiac antrum.

Predisposing Factors

Psychasthenia is believed to predispose to cardiospasm. Plummer and Vinson found that cardiospasm without dilation of the esophagus was associated in 100% of 24 cases of neuroses, while cardiospasm accompanied by dilation was present in only 5 of 91 neurotics. In Chevalier Jackson's group of 50 patients, the following exciting factors were given: nerves, excitement, worry, fright, anger, etc. - 40 cases; fatigue or overwork - 7 cases; hasty eating and improper mastication - 5 cases.

Symptoms

In the mild cases, the cardia periodically offers resistance, usually with discomfort or less often pain on swallowing and a sense that food sticks behind the sternum. In more severe cases, there are occasional stronger spasms with difficulty to force food through readily, and the patient has intermittent attacks of regurgitation during eating. Finally, in advanced cases, there is dilation and regurgitation which comes on regularly at intervals up to 36 hours after eating. Food in the dilated esophagus gives a sense of weight in the chest, sometimes with cyanosis and cold sweating. The regurgitated food is not sour. Gas may accumulate in the stomach without being able to escape, giving anginoid pain. Cardiospasm has been mistaken for angina pectoris, cholecystitis, neurasthenia, etc. In the beginning, the patient has difficulty only with heavy foods, then with semisolids, and finally with liquids. Coffee is especially troublesome. Ordinarily, patients find means to aid the passage of food such as taking deep breaths, taking much fluid with foods, standing while eating, or subsisting on fluids alone, preferably warm fluids.

Physical Findings and Diagnosis

There are so few physical findings that usually no change can be demonstrated on examination. A little dullness to the right of the sternum and absence of the second swallowing sound usually are the only signs. The condition is diagnosed on the history alone and is confirmed by the esophagoscope and x-ray examination. With the aid of barium, the x-ray picture is almost pathognomonic. The characteristic cigar or pear-shaped

dilation, larger than with carcinoma, appears with the hiatus opening from its left wall. The walls are smooth and fill evenly with barium, distinguishing the advanced disease from carcinoma. If the obstruction is not too marked, barium is seen passing in a fine stream into the stomach.

Esophagoscopy is of particular aid in determining the condition of the mucosa, which in advanced cases becomes grayish in appearance and may have superficial abrasions. Any considerable ulceration should suggest malignancy.

The passage of bougies is especially helpful, especially if done over a previously swallowed thread which has been allowed to become anchored in the intestines. Freeman considers this the most satisfactory way to differentiate between a simple spasmodic closure of the cardia and an organic stenosis, such as early carcinoma.

Complications and Sequelae

Starvation is an important complication. It may be the direct cause of death. Spontaneously, rupture and mediastinitis rarely occur. This is more often due to manipulation. Aspiration pneumonia, carcinoma of the sac, and ulcers of the esophagus or stomach have been recorded.

Soper reported 29 cases of cardiospasm. Of these, 7 had peptic ulcer and 3 had carcinoma of the stomach or esophagus.

Treatment

Medical treatment has been uniformly unsuccessful. The most satisfactory methods have depended on dilation of the cardia either through the esophagus or through a gastrostomy. Simple passage of bougies is inadequate. Various dilators are in use. Russel devised a rubber bag which could be distended with water after being placed in the cardia. Jackson performs repeated dilations from above with an air bag, together with medical supervision and a bland diet. Many German writers use mechanical dilators inserted through the esophagus, an effort being made to rupture the sphincter and leave the mucosa intact. Certain others advocate a diathermic probe.

Numerous operative measures have been used, chiefly abroad. Heller used an operation on the cardia similar to the Ramsted on the pylorus. Heyramsky advocated an esophago-gastrostomy. Mikulicz favored digital dilatation through a gastrostomy. Retrograde dilators have also been used.

Craig reported from the Mayo Clinic a cure of an otherwise intractable case by interruption of the sympathetics.

Prognosis

Untreated cases may die of starvation. The chief hazard of treatment is rupture of the esophagus. With hydrostatic dilators 75% are permanently cured (Wainwright), 65+% can be cured in one dilation (Shepherd), those who are not cured show recurrence in 4 weeks. It has been suggested that this is due to faulty placing of the dilators.

University of Minnesota cases - 7 in past 30 months.

Case	Age	Duration of Symptoms	Complicating Condition	Treatment	Outcome
1	34	3 mos.	Hypertension	0	?
2	31	1 yr.	0	4	Fatal mediastinitis
3	56	2 mo.	0	1	Cure
4	15	2 mo.	Bleeding congenital varis of esophagus	18 transfusions	Cure
5	33	8 yr.	0	1	Improved
6	21	10 mo.	0	2	Cure
7	65	40 yr.	Perforated peptic ulcer	1	Fatal peritonitis

Mayo Clinic - Vinson, 683 cases.

Treated by hydrostatic bag

Total number of cases	683	
Total treated by dilation	679	
Untreated	121	
Traced	562	
Considered cured		
Completely cured	188	)
		(-- 72.3%
Slight dysphagia	207	)
Moderate dysphagia	91	
Considerable dysphagia but some permanent benefit	26	
No permanent benefit	25	
Died while under observation		
Split at cardia	9	
Starvation before dilation could be done	2	

Treated by sound only

Total number of cases	41	
Untraced	6	
Traced	35	
Completely relieved	9	)
		(-- 54.3%
Slight dysphagia	10	)
Moderate dysphagia	5	
Considerable dysphagia	6	
No benefit	3	

Summary

1. Cardiospasm is a relatively uncommon disease although from larger clinics several hundred cases have been reported. In the Mayo Clinic, a total of 683 cases were seen in a period of twenty years.

2. The University of Minnesota Hospitals had 7 cases in the past 30 months.

3. The gross pathology consists of dilation of the lower two-thirds of the intrathoracic esophagus, accompanied by hypertrophy of the muscle apparently due to inorganic stenosis at or near the diaphragmatic hiatus. Not uncommonly, there are chronic inflammatory changes in the mucosa of the esophagus with thickening of the mucosa; ulceration of the esophagus has occasionally been reported.

4. Microscopically, the essential changes are hypertrophy of muscle and occasionally degenerating changes within the muscle such as fatty metamorphosis and hyaline degeneration. The nerve plexus shows degeneration evidenced by atrophy and infiltration of small round cells about the remnants of the ganglion cells.

5. It is conceded that the origin of the condition is primarily a disturbance in the peristalsis of the esophagus. Apparently, the cardia fails to relax when food reaches this portion of the tube.

6. Anatomically, there is said to be a normal thickening of the muscle about 5 cm. above the stomach and this is called the superior sphincter. At the esophageal hiatus, there is a second constriction and there is an inferior sphincter which apparently is made up of a ringlike fold of mucosa separating this portion of the esophagus from the stomach. This lower section of the esophagus is known as the cardiac antrum.

7. Congenital malformation, infection, irritation from poorly chewed food, irritation from local diseases, hypertrophy of the diaphragmatic muscle and nerve imbalance are the suggested causes of the spasm.

8. The theory which receives greatest support is the theory of nerve imbalance. The vagus nerve maintains the tone of the muscle and atrophy or section of this nerve produces dilation of the esophagus. Section of vagi in cases of cardiospasm apparently makes the condition worse. Section of vagi in dogs has been said to produce the gross and microscopic picture of cardiospasm. In one case, sympathetic ganglionectomy has resulted in complete cure.

9. In one series of 24 neurotics, all manifested some degree of cardiospasm but there was no dilation above the spastic area. In another group of 50 patients with cardiospasm, 40 stated that worry, fright, anger, excitement, nervousness, etc. made the condition worse or brought on an attack.

10. In the mild cases, there is only periodic resistance to swallowing with some sensation and discomfort in the chest. More severe cases may show occasional severe seizures in which food cannot be swallowed and there may be intermittent attacks of regurgitation. In the well advanced cases, the inability to swallow food may progress to such an extent that even liquids cannot be swallowed. The regurgitation may take place immediately after eating or may be delayed as long as 36 hours.

11. In some cases, the symptoms may suggest angina pectoris, cholecystitis, gastric disturbances, neurasthenia, etc.

12. As the corollary to this differential diagnosis, it should be added that not all the symptoms may be due to cardiospasm in that diverticulae, ulcers and even carcinoma may be associated with the cardiospasm (may be causative agent?).

13. There are so few physical findings that demonstration of the lesion on examination usually is difficult. There may be some increase in mediastinal dullness.

14. The diagnosis should be made on the history and confirmed by roentgenological examination.

15. The roentgenological criteria are very characteristic to a cigar or pear-shaped dilation, usually larger than with carcinoma, with its hiatus opening from its left side into the stomach. The walls are smooth, filled evenly with barium and when the obstruction is not too marked the barium may be seen in the passages.

16. Esophagoscopy is of particular value because the condition of the mucosa may be determined and the possibility of ulceration and malignancy can be ruled out.

17. Bougies are used to distinguish between simple spasmodic closure, organic stenosis and carcinoma.

18. Starvation is the most important complication in untreated cases. Other causes of death may be aspiration pneumonia and ulceration.

19. The treatment of this condition is not without its risk. Traumatic rupture of the esophagus with mediastinitis is the chief danger.

20. Medical treatment has been uniformly unsuccessful.

21. Operative measures have been used chiefly abroad, plastic operation similar to Ramstedt, and various types of esophagogastrostomy have been tried.

22. One cure has been reported following sympathetic ganglionectomy.

23. Simple passage of bougies is inadequate. Repeated dilation from above using either air or water bags is the method of choice. This is supported with medical management and a bland diet.

24. It is said that 70% may be cured in one sitting and 75% are permanently cured.

25. When recurrence takes place, it is usually present within 4 weeks and it is said to be due to faulty placing of dilator.

26. In the University of Minnesota series, there were 7 cases, 2 of which died, one from a fatal mediastinitis, the other from peritonitis secondary to rupture of a duodenal ulcer. One case has not been followed, three have been cured, and one has improved.

27. In the Mayo Clinic group of 683 cases, 72% were cured or had only slight dysphagia following treatment with hydrostatic bag.

28. The result of treatment by sound only hielded 54% cures.

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II. CASE REPORT

CARDIOSPASM. PERITONITIS.

Case is white male, 74 years of age, admitted to University of Minnesota Hospitals 7-11-35 and expired 7-19-35 (8 days).

40-yr. history

1931 - Has had trouble with swallowing from time to time for past 40 years. Difficulty progressed so it has not become necessary to pass bougies to dilate the obstruction.

Dilations - Partial Success

1934 - Has had about 10 dilations which have been fairly successful. In the past few months, the procedure has given less and less relief. Suffered attacks of bronchitis which made the difficulty in swallowing much worse.

Worse - Regurgitation - Weight Loss

7-11-35 - Admitted. Progressive increase in difficulty in swallowing. Cannot swallow solid foods. When he lies down and puts his head to the side, he regurgitates liquid which he had previously swallowed. Lost about 20 lbs. and felt weak and short of breath. Physical examination: Chest - emphysematous, practically no excursion in breathing; heart tones, distant. Abdomen - extremely emaciated; liver and spleen not palpable; some dilated veins over abdomen. Extremities - negative. Kernig sign - negative. Laboratory: Urine - negative. Blood - hemoglobin 100%, leucocytes 11,900, neutrophiles 82%, lymphocytes 15%. Progress: Attempt made to pass thread upon which a bougie could be guided but this was unsuccessful.

Esophagoscopy

7-15-35 - Under local anesthesia, attempt was made to esophagoscope patient. Esophagus enormously dilated and filled with fluid. Partial evacuation performed. Continued on intravenous fluid. Esophagus evacuated daily; as much as 1000 cc. could be recovered.

Dilated - Sudden Death

7-19-35 - Under local anesthesia, esophagoscope again passed. Spasm at lower end of esophagus would only permit passage of a #30 sound. In afternoon, patient seemed very weak. Pulse rapid. Slight increase in temperature. 8:25 P.M. Suddenly collapsed. Expired.

Autopsy

Emaciation

Body is poorly nourished, white male, 74 years of age, measuring 164 cm. in length and weighing approximately 110 lbs. Development is of small type. Rigor is present. Hypostasis is purplish and posterior. Edema of ankles. No cyanosis. Pupils are equal, each measuring about 3 mm. in diameter. No special marks about body except for moles on left chest and neck and about ankles.

Peritonitis (old) - Pneumoperitoneum

Peritoneal Cavity shows presence of gas and purulent exudate throughout the cavity. This exudate in many places seems to become organized, especially under the diaphragm where it is fibrous and shaggy. Appendix shows no change.

Pleural Cavities and Pericardial Sac contain no adhesions or excess fluid.

1+ Coronary Sclerosis

Heart weighs 290 grams, is small and flabby and somewhat brownish. No hypertrophy or dilation. Musculature shows no softening or fibrosis. Endocardium and valves are smooth. Root of Aorta shows atheromatous plaques. Coronaries show few small, yellow beadings. In major trunks, there is no significant obstruction.

Emphysema

Right Lung weighs 500 grams, Left 425. There is no bronchopneumonia. Bronchi contain considerable amount of mucoid material. Both lungs are somewhat emphysematous.

Spleen weighs 125 grams, shows no change.

Liver weighs 1450 grams. Markings are well retained. No abscesses, tumors or cirrhosis.

Gallbladder wall is thin. Mucosa is smooth. No stones.

Gastro-Intestinal Tract

The esophagus is enormously dilated. Stomach and esophagus are fixed by filling it with formalin solution, 1,600 cc. being poured in. The major part of this remaining within the esophagus so that it is estimated that the esophagus easily holds between 1000 and 1400 cc. of fluid. The average diameter of the esophagus is approximately 10 cm. The esophagus is egg-shaped and bulges into both pleural cavities. There is no evidence of perforation of the esophagus on its external surface. There is no digestion and no exudate on the outer side. The muscle of the esophagus appears thick, averaging about 6 to 8 mm. in thickness. The dilation is uniform, beginning just below the thyroid cartilage extending down to the diaphragm and ending sharply as the esophagus passes thru the diaphragm. On the inner surface of the esophagus, there are several ulcers, most of these being round and punched-out, others stellate and irregular. One of these penetrates through the full thickness of the esophagus and the base is made up of the adventitious tissue outside of the muscle layer. There has been no penetration or spread outside of the process. This ulcer is approximately at the junction of the upper and lower two-thirds. All of the ulcerations are within the lower two-thirds of the esophagus. As the esophagus passed through the diaphragm, it shows no gross change other than the small size of the lumen in comparison to the dilation above. There is no tumor or stricture. The esophagus can readily be spread with the fingers at this point.

The stomach is small. The mucosa is smooth. No ulcers or tumors. The pylorus shows no change. In the first part of the duodenum, there is an ulcer on the superior surface of the duodenum which has penetrated through the full thickness of the wall and perforated into the free peritoneal cavity. In the second part of the duodenum there is another ulcer which appears to be more chronic, harder and stellate in outline. The base of this ulcer has penetrated into the head of the pancreas. The remainder of the gastro-intestinal tract shows no change other than an empty collapsed state and a few shreds of mucus in the small and large bowel. No diverticulae or other ulcers.

Pancreas is small and soft.

Kidneys each weigh 120 grams and show

no significant change. Bladder is normal.

Adrenals are well developed.
Prostate is small and soft.

Diagnosis

1. Cardiospasm.
2. Dilation and hypertrophy of esophagus.
3. Multiple ulcers of esophagus.
4. Multiple duodenal ulcers.
5. Perforation of duodenal ulcers.
6. Peritonitis.
7. Emphysema.

III. MOVIE

Title: The Peacock Throne

Released by: Monogram Pictures.

IV. LAST WEEK

Date: October 31, 1935

Place: Recreation Room, Nurses' Hall

Time: 12:15 - 1:25

Program: Movie (Community Chest)
Biliary Fistula

Present: 101

Discussion: C. N. Borman
L. G. Rigler
O. H. Wangensteen

Gertrude Gunn,
Librarian

V. GOSSIP

The Minnesota State Medical Association will meet in Rochester in 1936. At one time the meetings were confined to St. Paul, Minneapolis and Duluth, with an occasional trip to a smaller community. In the past few years, however, Rochester has joined the list of the regulars. These meetings at one time very small, compact, and orderly, now take on the appearance of a national meeting with large exhibit sections, lectureships, movies, demonstrations, etc. The meetings now draw an attendance of over 2000.....William Thomas Peyton finds but little lure in the summertime in Minnesota but when Fall comes around his vacation time has arrived. He left last week on his annual jaunt with dog and gun and when he returns he finds his friends waiting for his yarns. In one of his hunting trips a few years ago there is the story of the bear killed by one of the members of the party. This mighty huntsman, now a prominent surgeon in Duluth, was much disturbed to get a letter from the owner of the bear asking for $100.00 as the little children had not been happy since their playmate was taken away.....There was a very pleasant afterglow from the short course, although some of the criticisms about the arrangement are justified. We have in our group certain men who are able to present to students of all degrees of intellectual advancement interesting and instructive clinics and lectures. We have others who feel that no preparation is necessary, or that they can give them some old stuff they have had on hand for a long time. One of our outstanding examples of a man who presents his material well is Hobart Reimann. He differs from the rest of us in that no one has any misconception as to what he thinks. I have heard many favorable comments about his manner of presentation, even from the men who most violently disagree with him. Off the job Internist Reimann doffs his dignity of the day to become an entertaining socialite by night. Few men in our group have the knowledge of the games, stunts, trick costumes, playlets, etc., and the ability to put them over, as one Hobart Reimann.....Captain Glenn Seidel, on the football program last night, was most careful to explain that it was not Beise

tackling Simmons which caused the damage, but rather Simmons tackling Beise, but it is a matter of small difference as the result is the same.....Much credit is due the interns, fellows, and staff men for their assistance in preparing the programs for this year. We have had more favorable comment about our meetings than ever before, and certainly the interest is running higher. You may be interested to know that we now have programs prepared well up into the year, but there are many men on the staff who have something to offer the rest of us, who have not yet signed on the dotted line. Rudolph Koucky deserves credit for developing this plan of presentation. .....Author Harold Shelly Diehl (Healthful Living) is learning that there are certain members of your reading public who violently disagree with your opinions no matter how innocent they may be..... Freshman Clinic is still packing them in. This "no credit, no attendance, no examination" course has a waiting audience long before the appointed time. An opportunity to present the fundamental facts of medicine and medical practice is apparent to all who have seen the interested audience. It has been found that our deepest impressions come from the earliest stages of our medical education. The amazing thing about the audience is its ability to handle difficult diagnostic problems with apparent ease. Some of our students actually have the ability to reason. Perhaps they lose it later on, or we do not encourage its development.....Dr. Morris Fishbein was most pleased with the staff meeting he attended. Felt that we were doing a good piece of work and dignified our sheet by asking, would it be sent to him? After every visit the local "Docs" always scan "Tonics and Sedatives" for some sort of mention or impression of the trip. It is certainly the most widely read feature in the Journal.