CONGENITAL PARTIAL PERICARDIAL DEFECT WITH HERNIATION OF THE LEFT ATRIAL APPENDAGE

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INTRODUCTION

Congenital defects of the pericardium are considered rare. Until 1979, the reported cases were only about 200.1 Total absence of the left pericardium is the most common defect2 and less common is a partial defect of the left pericardium. Other types, i.e. isolated right-sided defects, total pericardial absence and diaphragmatic pericardial defects are very rare.3

Before the last decade, the diagnosis of partial defects of the left pericardium was very difficult and sometimes made only during surgical intervention or post-mortem examination. More recently however, the use of angiocardiography and induced left pneumothorax have made diagnosis of this kind of pericardial defect relatively easy. In the present case, the only finding was the prominence of the left hilum on radiography and the correct diagnosis was made through angiocardiography and left pneumothorax.

CASE REPORT

A 40 year old woman was admitted to the hospital because of episodes of coughing of three months duration. There were no obvious precipitating factors and no history of cardiovascular disease could be obtained. On physical examination she was well developed, blood pressure was 120/70, pulse rate was 75, and temperature was 37°C. Pulmonary and cardiac auscultation revealed no abnormalities. There was no cyanosis and the extremities were normal. A chest roentgenogram disclosed a normal-sized heart with a prominent left hilum (Fig.1) Chest tomography was performed and the diagnosis of masses or tumors was ruled out. With a presumptive diagnosis of pericardial defect, a right-heart catheterisation and pulmonary artery angiography were performed. Pressures were all within the normal range. On angiocardiography, the size and shape of the pulmonary artery were normal, but the levo-phase showed the left atrial appendage to occupy and make up the abnormal left hilar shadow (Fig.2). The degree of herniation varied with the contraction of the left atrium. After injection of 500cc carbon dioxide into the left pleural cavity, the left lateral decubitus view showed a very thin layer of carbon dioxide in the right pericardial area, which confirmed the defect of the left pericardium that allowed the entrance of the gas from the left pleural space through the defect into the pericardium. The patient was managed conservatively.

DISCUSSION

Partial defect of the left pericardium has been reported in 35% of all congenital pericardial defects.1 In the series studied by Ahn, et al,3 out of 55 cases of partial left pericardial defect, there were 29 associated congenital anomalies, from which 13 cases presented a cardiac malformation. The most common associated cardiovascular congenital anomalies are patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot and atrial septal defect.1 Fifty percent of partial left defects are complicated by herniation of the left atrial appendage.1

The embryological cause of partial left pericardial defect is thought to be premature atrophy of the left duct of Cuvier which supplies nourishment to the pleuro-pericardial membrane.4 Clinically, isolated cases may be asymptomatic or the patient may occasionally complain of episodes of tachycardia5 or chest...
Congenital Partial Pericardial Defect

Figure 1. Chest x-ray showing a normal-sized heart with a prominent left hilum.

pains. Other symptoms such as dizziness and syncope are noted as well. Physical findings are very rare in the isolated forms. Some investigators could find a pulmonary ejection murmur and even splitting of the second heart sound. Roentgenographic findings in total absence of the pericardium are characteristic and include ashift of the heart to the left, and flattening and stretching of the inferior portion of the cardiac silhouette over the dome of the left hemidiaphragm. Left partial defect however can only be suspected by radiographic findings of a prominent left hilum if there is herniation of the left atrial appendage through the defect. Usual cases without herniation present no roentgenographic abnormalities.

The most decisive procedures for diagnosis are angiography and artificial left pneumothorax. Angiography will demonstrate that the abnormal portion of the left heart contour is occupied by the left atrial appendage and in some cases may even reveal that the silhouette of the left appendage stretches beyond the confines of the pericardium. Artificial left pneumothorax will permit the final diagnosis and differentiate between idiopathic dilatation of the left atrial appendage and partial pericardial defect. This method consists of injecting 400-500 ml of CO₂ into the left pleural space. In the left lateral decubitus position, one can observe a very thin layer of air between the right heart contour and the right lung field which is characteristic of this condition. Once the diagnosis is established, some authors do not recommend surgical intervention until the emergence of chest pain or dysrhythmias. Others recommend surgical intervention even for asymptomatic patients, because of potential catastrophic herniation and strangulation, although there have been no reported deaths from strangulation of the left atrium or left appendage. There have been three reported deaths from strangulation of the left ventricle due to partial defect of the left pericardium. Nevertheless, surgical treatment including atrial appendectomy, pericardial resection, and pericardioplasty by Teflon patches or fascia lata may protect the heart from herniation and from adjacent pulmonary infections. It seems also that surgical correction of the defect may offer the heart optimal position for its function and thus prevent the occurrence of cardiac irritability.

Summary

A 40 year old woman is presented, who was complaining of intermittent short episodes of coughing. Chest radiography revealed an abnormal left hilum. The diagnosis of partial left pericardial defect with herniation of the left atrial appendage has been established by angiography and artificial left pneumothorax. A brief review of the literature regarding signs and symptoms, roentgenographic findings, angiography, induced pneumothorax, and management is presented.

REFERENCES
2. Lind T.A., Pitt M.J., Groves B.M., White J.E. and Quinn E: The

Editorial Comment

This article is an interesting presentation of a rare pericardial anomaly. It must be noted however that in approaching this type of anomaly, most authors do not recommend artificial pneumothorax as a diagnostic procedure.

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URACHAL DIVERTICULUM WITH
ALTERNATING SINUS

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ABSTRACT

Urachal anomalies are exceptionally rare. They can present in different
ways in childhood as well as in adults. We present an interesting case of
vesico-urachal diverticulum with alternating sinus. The interesting finding in
this patient was the inversion and disappearance of the umbilicus at the
termination of voiding.

CASE REPORT

A 16 year old male was referred to us due to
occasional egress of urine from his umbilicus.
He would also complain of an inward pulling
sensation and disappearance of his umbilicus at the
end of urination. No other urinary symptom was
present. The patient stated that he had been passing
urine often through his umbilicus during his infancy
and childhood. Routine physical examination re­
vealed no significant findings. We observed the
patient passing urine *per urethra* and noticed the
umbilicus disappearing and assuming a pin-hole
appearance. He seemed to have a normal urinary
stream. The intravenous urogram was normal. A
voiding cystourethrogram was done. After filling the
bladder to its highest capacity with contrast material
we were able to demonstrate a diverticular out­
pouching at the dome of the bladder extending
toward the umbilicus (Fig. 1). No distal obstruction
was seen (Fig. 2). We were unable to reveal the
connection between this urachal diverticulum and the
outside through the umbilicus, which clinically has
existed since the patient’s birth. Cystoscopy revealed
a normal urethra and bladder neck. The bladder was
not trabeculated. A urachal diverticular opening was
noticeable at the anterior portion of the bladder
dome in the midline. The patient underwent
surgery. The urachal diverticulum was removed
completely from the dome of the bladder up to the
umbilicus via a lower abdominal midline incision. The
bladder wall at the urachal diverticular site was
repaired in two layers. The post-operative course was
uneventful. The umbilical inversion sign noticed
during the terminal phase of voiding preoperatively
disappeared after surgery.

Fig. 1. A vesicourachal diverticulum demonstrated in a cystogram
of a young male.
DISCUSSION

Urachal anomalies are exceptionally rare occurrences that may manifest themselves in later life as well as in childhood. Blichert-Toft and Nielsena were only able to collect 315 cases of patent urachus in the literature (1971) and only three cases were seen in more than one million admissions at the Charity Hospital in New Orleans (Nix, et al.). The urachus is a tubular three-layered structure that courses upward between peritoneum and transversalis fascia from the anterior dome of the bladder toward the umbilicus. Failure of the urachal lumen to close will result in one of the several presentations of urachal anomalies. Our patient had a vesico-urachal diverticulum with an alternating sinus draining intermittently via the umbilical route. Clinically these patients may present in different ways. Patent urachus usually presents soon after birth but not until the cord separates from the umbilicus. A distal obstruction may be present which can be visualized or ruled out by voiding cystourethrography.

Some patients may present with lower abdominal mass in the midline (urachal cyst) and others as omphalitis. Chronic urinary tract infection and stone formation may be the presenting symptoms in longstanding cases of large urachal diverticuli. In many cases there are no symptoms and the anomaly may be found incidentally during unrelated studies. The interesting finding in our patient was the inversion and complete disappearance of the umbilicus in the final stages of voiding. In fact, detrusor muscle contraction pulled the umbilicus inward through the urachal attachment.

REFERENCES