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CONGENITAL PARTIAL PERICARDIAL DEFECT WITH HERNIATION OF THE LEFT ATRIAL APPENDAGE

M. RADPOUR, B. KHADIVI, AND F. MAHDAVIAN

From the Department of Internal Medicine and Department of Surgery, Mashad University of Medical Sciences, Mashad, Islamic Republic of Iran

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 defect[2] 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.[3]

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 tachycardia[5] or chest

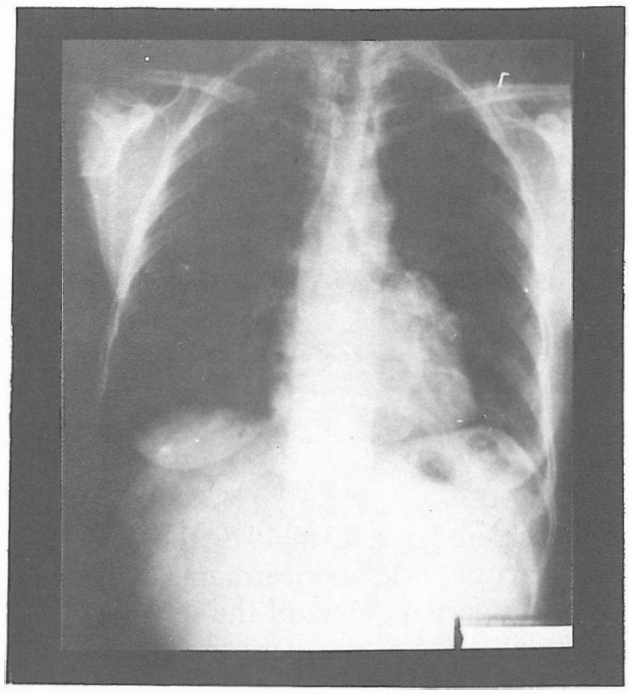

Figure 1. Chest x-ray showing a normal-sized heart with a prominant left hilum.

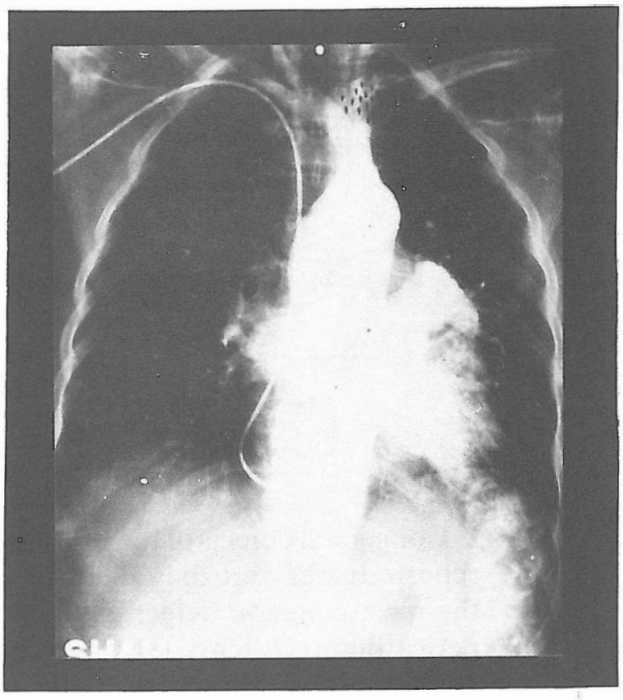

Figure 2. Angiocardiography reveals that the left atrial appendage makes up the abnormal left hilar shadow.

pain.[6] Other symptoms such as dizziness and syncopes are noted as well.[1] 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.[7] Roentgenographic findings in total absence of the pericardium are characteristic and include a shift 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.[8]

The most decisive procedures for diagnosis are angiocardiography and artificial left pneumothorax. Angiocardiography will demonstrate that the abnormal portion of the left heart contour is occupied by the left atrial appendage[9] and in some cases may even reveal that the silhouette of the left appendage stretches beyond the confines of the pericardium.[6] 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 CO_2 into the left pleural space. In the left lateral decubitus projection, 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.[9,10] Once the diagnosis is established, some authors do not recommend surgical intervention until the emergence of chest pain or dysrythmias.[3] Others recommend surgical intervention even for asymptomatic patients, because of potential catastrophic herniation and strangulation,[4,5] although there have been no reported deaths from strangulation of the left atrium or left appendage. There have been three reported deaths from stangulation of the left ventricle due to partial defect of the left pericardium.[11,12] 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.[5] 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.[5]

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 angiocardiography and artificial left pneumothorax. A brif review of the literature regarding signs and symptoms, roentgenographic findings, angiocardiography, induced pneumothorax, and management is presented.

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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.

Hassan Arefi, M.D.

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URACHAL DIVERTICULUM WITH ALTERNATING SINUS

NASSER SIMFOROOSH

From the Department of Urology, Shahid Labbaf i-Nejad Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Islamic Repubic of Iran

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 revealed 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 outpouching at the dome of the bladder extending toward the umbilcius (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.

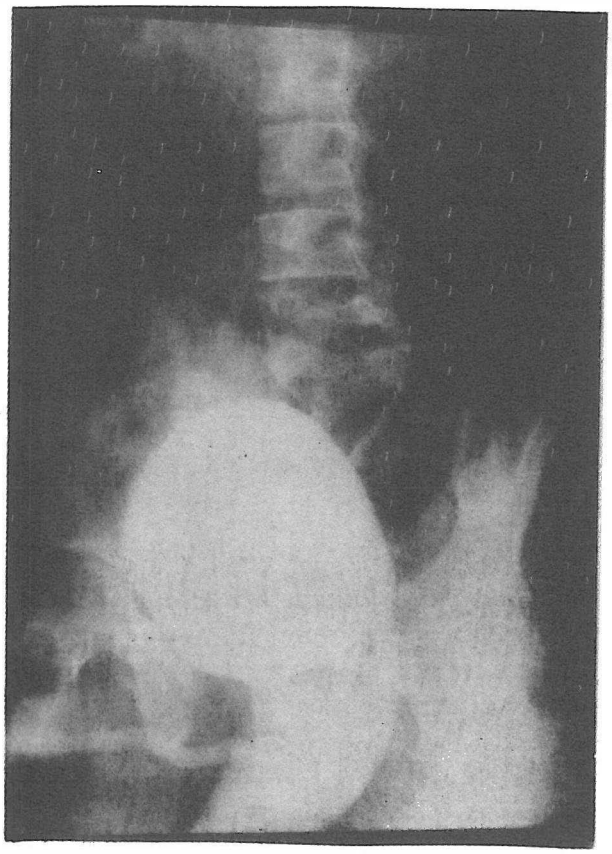

Fig. 2. Voiding cystourethrogram in a patient with a urachal diverticulum and alternating sinus. No obstruction is seen in the bladder neck or urethra, and the bladder does not seem trabeculated.

DISCUSSION

Urachal anomalies are exceptionally rare occurrences that may manifest themselves in later life as well as in childhood.[1] Blichert-Toft and Nielson[2] 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).[3] 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 long-standing 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.

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