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Case report

Pericardial Cyst in a Neonate

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Pericardial cyst is a rare entity, often discovered incidentally on routine chest radiographs. Some studies reported an incidence of about 1:100.000.[1-5] Symptomatic cysts presenting in the neonatal period are very rare.[2] Most of the patients are asymptomatic, but can cause symptoms when they undergo complications or on account of their large size.[1] We describe the case of a neonate presenting with tachypneau where a pericardial cyst was diagnosed with echocardiography and computer tomography, the cyst was successfully resected with open heart surgery.

Case Illustration

A neonate was referred to The National Cardiovascular Center Harapan Kita for the evaluation of tachypneau, feeding difficulty and an abnormal chest radiograph. He was born at 35 weeks gestation by cesarean section due to early rupture of the membrane to a G2P1 30 years old mother. The mother had routine antenatal care at a nearby clinic, and in 34 weeks gestation an antenatal ultrasonography scan showed a cystic thoracic mass. Soon after birth the symptoms started. No history of fever or signs of infection. Physical examination showed no pathologic findings, no cyanosis, normal heart sounds, no murmur, and no pathologic auscultation of the lungs. Chest x-ray showed enlarged heart silhouette especially to left superior (**Figure 1**). On echocardiography a multilloculated pericardial cyst at the left anterior pericardium was detected (**Figure 2-3**), with moderate pericardial effusion, no signs of cardiac tamponade. The heart and great vessels were otherwise normal. A computer tomography supported these findings (Fig. 4) showing a hypodens (17 HU) mass suggestive of fluid with clear border at the upper left pericardium to the left apical pleura.

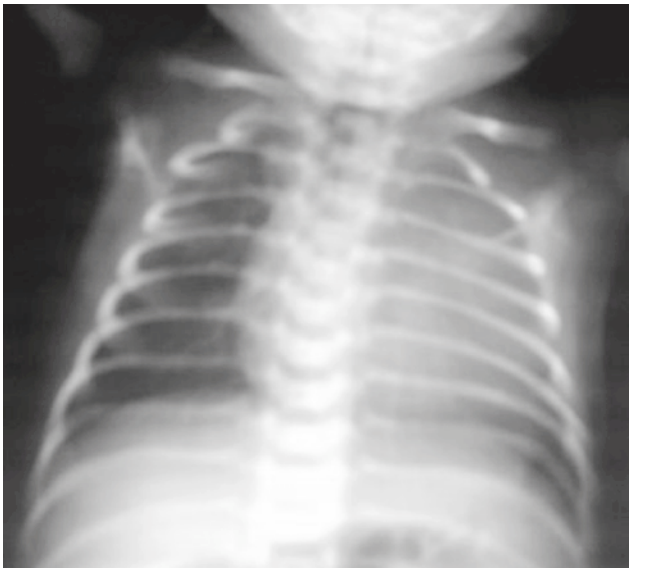

Figure 1. Chest x-ray showed enlarged left superior heart silhouette.

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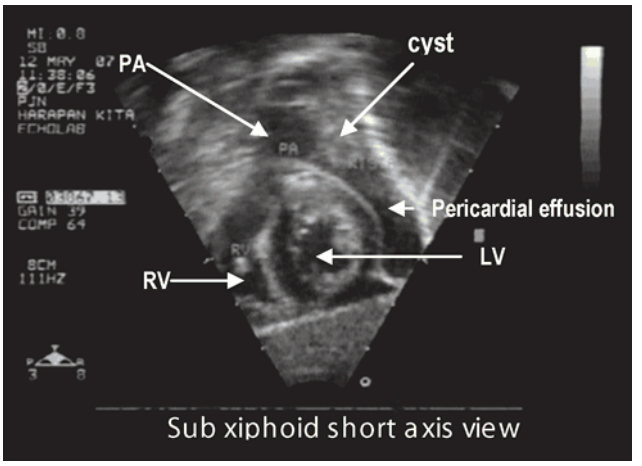

Figure 2. Echocardiography Subxiphoid short axis view showed pericardial effusion and cyst in the pericardial cavity

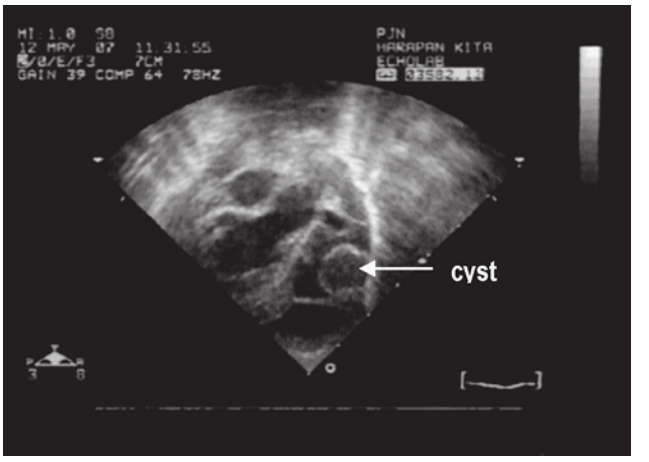

Figure 3. Echocardiography Subxiphoid long axis view showed cyst in the pericardial cavity

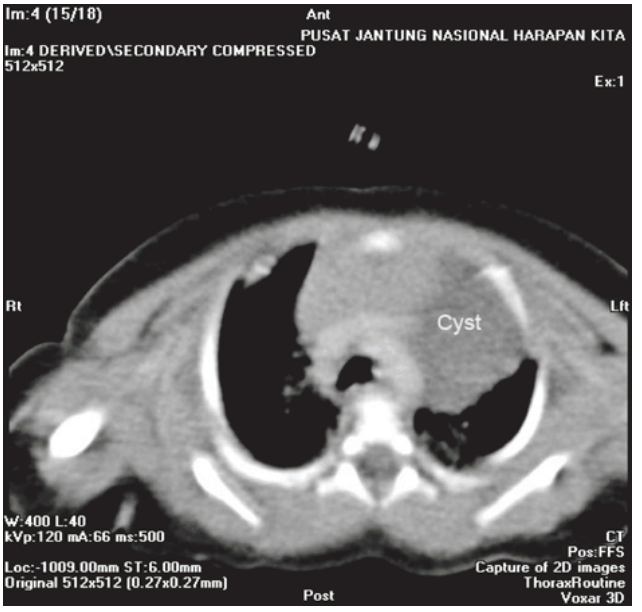

Figure 4. Computer tomography scan showed a hypodens (17 HU) mass, suggestive of fluid with clear border at the upper left pericardium to the left apical pleura.

Since the symptoms were progressive, the baby underwent elective open surgery for cyst resection. Intra-operative findings supported the diagnosis of multiloculated pericardial cyst: it measured 6.0 cm x 2.2 cm x 1.5 cm and 3.0 cm x 1.4 cm x 0.7 cm. The procedure and post operative course was uneventful. Histologically, the findings were consistent with pericardial cyst. Short term follow up showed a good clinical result and no residual cyst on echocardiography.

Discussion

Pericardial cysts are an uncommon benign congenital anomaly in the mediastinum. They represent 6% of mediastinal masses and 33% of mediastinal cyst.[6] In the mediastinum 61% of presenting masses are cystic. Pericardial and bronchogenic cyst share the second most common etiology of mediastinal masses after lymphoma.[5,6] They are thought to result from failure of fetal lacunae to coalesce into the pericardial coelom.[7] Though congenital in origin, they usually present in the older population.[8] There are very few reports of pericardial cysts in the neonate.[9] It is possible to detect them antenatally as early as in mid second trimester.[1]

These cysts are usually asymptomatic, but become apparent on account of their size or after undergoing some complication. The presentations include respiratory distress, cardiac tamponade, heart failure, pain due to torsion and rupture of the cyst causing pleural effusion and pericardial effusion.[5,8] Diagnosis is based on radiology. They could be incidentally picked up on chest X-ray. For confirmation a CT scan and 2-D Echo are sufficient.[10,11] No further invasive investigation is necessary. However, only a histo-pathological examination of the tumor is conclusive. The characteristic CT findings are a cyst occurring in the cardiophrenic angle (70% on the right)[1-5] with smooth walls and near-water attenuation of the fluid contents. They may also occur in other atypical locations.[12] Our case is therefore unusual, because the cyst presented in a neonate and in an atypical mediastinal location.

The differential diagnoses includes a number of conditions : Teratoma, Hemangioma, Fibroma, Bronchogenic cyst, esophageal duplication cyst, Extralobar sequestration, Lymphangioma, Lipoma, Neurogenic tumor, Sarcoma and Abscess.[8,10]

The management of pericardial cyst includes observation, percutaneous drainage and operative resection.[5] This depends upon the presentation and the available resources.[1] Observation is possible with repeated CT scans on asymptomatic individuals. However, there is little information regarding the safety and appropriate length of observation.[5] In the acute setting it may be necessary to aspirate the cyst to control heart failure or to improve respiratory status.[1] Emergency thoracotomy has also been reported for excision of the mass.[9]

In stable patients, the treatment options could be either surgery or aspiration. Aspiration alone is a viable option if the cyst fluid is clear and there is no doubt of the diagnosis. These patients need long term follow up and serial CT scans to determine any recurrence.[1] One literature review reported that one third of patients had recurrence after percutaneous drainage at three years.[5]

Operative intervention is used as an adjunct to the aspiration or as a primary treatment modality. It is usually performed when there is a doubt about the diagnosis or recurrence.[1] Operative intervention has become more attractive with the advent of video-assisted thoracic surgery (VATS). The obvious advantages of this procedure is that it is minimally invasive and involves less morbidity than open surgery.[2] These are partly offset by the associated difficulty that may be encountered during dissection on account of the position, surrounding inflammation, bleeding and suturing. The difficult sites include those adjacent to the bronchus, the esophagus and the subcarinal area. In these difficult situations as in this case the open method is preferable.[1] Open surgery does not require special equipment. It is especially indicated in complicated and emergency situations. While the morbidity and mortality of pericardial cyst are unknown, surgery has demonstrated as the only definitive cure.[5]

Conclusion

Most pericardial cysts are asymptomatic, located in the right cardiophrenic angle and detected incidentally during routine chest films. Symptoms and serious complications such as dyspnea, cough, respiratory distress, chest pain and cardiac tamponade can occur as the result of an expanding lesion on vital adjacent structures. The diagnosis of pericardial cysts can be done prenatally using ultrasonography. Once suspected the diagnosis is established by noninvasive studies such as echocardiography and CT scans. Management of pericardial cysts is surgical resection whenever possible. The objective of removal the lesion is elimination of the tumorous mass, relieve of symptoms and allowance of histological examination. Approach can be open or video-assisted thoracoscopic surgery. Prognosis is excellent in most cases.

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