Chylopericardium presenting as cardiac tamponade secondary to mediastinal lymphangioma

MM Alsmady, MA Aladaileh, K Al-Zaben, MM Saleem, O Alimoglu

University of Jordan, Amman, Jordan

ABSTRACT

Mediastinal lymphangioma is a rare entity and chylopericardium is a rare form of pericardial effusion. We report a case of acute chylous cardiac tamponade due to a cervicomediastinal lymphangioma in a one-year-old boy. A chest x-ray revealed marked cardiac enlargement and echocardiography showed massive pericardial effusion. Emergency surgery was performed whereby a pericardial window was created, followed by excision of the lymphangioma.

KEYWORDS

Cystic lymphangioma – Cardiac tamponade – Chylopericardium

Accepted 17 April 2016

CORRESPONDENCE TO

Moaath Alsmady, E: moaathalsmady@yahoo.com

Described initially by Redenbacker in 1828, lymphangioma is considered one of several mediastinal and pulmonary lymphatic diseases.¹ It is formed by early sequestration of the lymphatic tissues, which arise from a primitive jugular lymph sac during embryogenesis.

There is no consensus regarding the actual incidence of lymphangiomas in the mediastinum but they are estimated to constitute 0.01–4.5% of mediastinal masses.¹ They are more common in the paediatric setting and are frequently located in the neck. We report a rare case of a lymphangioma with acute cardiac tamponade due to chylopericardium requiring surgery.

Case history

A one-year-old boy was admitted to our university hospital with neck swelling. Physical examination revealed tachycardia and hypotension while a chest x-ray revealed marked cardiac enlargement (Fig 1) and transthoracic echocardiography demonstrated massive pericardial effusion. Computed tomography (CT) showed a large cervical multicystic mass with a mediastinal extension and pericardial involvement (Figs 2 and 3).

Emergency surgery was carried out owing to progressively increasing pericardial effusion and the patient's worsening haemodynamic state. The pericardium was distended and 400ml of chyle was evacuated by means of a pericardial window. The cystic mass in the anterior mediastinum was excised. Pathological examination of the obtained specimens confirmed the diagnosis of lymphangioma. Postoperatively, a persistent chylothorax developed. This did not resolve with conservative management and reoperation was eventually required for ligation of the

thoracic duct through a right posterolateral thoracotomy. There were no clinical or radiological signs of recurrence during the next three years.

Discussion

Lymphangiomas are rare and usually benign. It is thought they originate from abnormal lymphatic system development. They are very slow growing neoplasms and are usually only found in one organ but occasionally, several organs in one region are involved. In the thorax, involvement of the mediastinum, lung, heart, pleura, pericardium, ribs and vertebrae may be discovered in the same individual. Histologically, they are made up of cysts lined with endothelium; these can vary in diameter from a fraction of a millimetre to several centimetres.²

It may be more appropriate to replace the term 'lymphangioma' with 'lymphatic malformation', as suggested in 2014 by the International Society for the Study of Vascular Anomalies (ISSVA).³ According to the ISSVA's widely used classification for vascular anomalies, tumours and malformations vary histologically, and include many different types. Lymphangiomas have traditionally been categorised into three types depending on the size of the lymphatic channels. The smallest type is known as capillary lymphangioma, followed by cavernous lymphangioma and finally, cystic lymphangioma (hygroma). Cystic lymphatic malformations are the most common. The mediastinal lesion in our patient was a mixed cystic lymphatic malformation as some of the constituent cysts were greater than 2cm in size while others were smaller.

Lymphangiomas are most often located in the neck. Approximately 1% are located in the thoracic area with a

Figure 1 Chest x-ray showing cardiomegaly

predilection for the anterior part of the mediastinum. Mediastinal lymphangiomas are usually extensions of cervical lymphangiomas (as in our patient); 10% of cervical lymphangiomas extend into the mediastinum.⁴

On radiography, lymphangiomas appear as well defined oval or round lesions. Chyle may sometimes be present as a pleural effusion. On CT, they appear as a smooth, lobulated mass, which may compress the adjacent mediastinal structures. Lymphangiomas usually have a homogeneous appearance and low attenuation similar to that of water but they can have higher attenuation if they consist of a combination of fluid, fat and solid tissue. It is rare to see calcification on CT. Lymphangiomas can be unilocular or multilocular with thin septations.⁴

Surgical excision is the standard treatment for lymphangiomas. For patients in whom the mass is too big or if vital structures are encased by the tumour, complete surgical resection may not be possible. In such circumstances, derooing the mass is mandatory and most of the wall should be excised.¹

Many reports in the literature show alternative treatments for managing patients when surgery is not feasible. These include laser therapy, radiotherapy, chemotherapy, sclerosing agents and observation (especially for lymphangiomas that are mostly located in the neck).¹

Lymphangiomas have a potential risk of recurrence, especially if there has been incomplete resection. Extensive resection is therefore recommended. The prognosis is extremely favourable following surgery although regular postoperative follow-up review is highly recommended.

Figure 2 Computed tomography of the neck showing cystic mass on the right side

Figure 3 Computed tomography of the chest showing huge pericardial effusion

Conclusions

Mediastinal and cervicomedastinal lymphangiomas with chylopericardium are rare conditions. There are relatively few published reports of chylous cardiac tamponade. Early creation of a pericardial window and surgical resection of the lymphatic malformation are warranted.

References

1. Park JG, Aubry MC, Godfrey JA, Midthun DE. Mediastinal lymphangioma: Mayo Clinic experience of 25 cases. *Mayo Clin Proc* 2006; **81**: 1,197–1,203.
2. Rostom AY. Treatment of thoracic lymphangiomatosis. *Arch Dis Child* 2000; **83**: 138–139.
3. Dasgupta R, Fishman SJ. ISSVA classification. *Semin Pediatr Surg* 2014; **23**: 158–161.
4. Jeung MY, Gasser B, Gangi A *et al.* Imaging of cystic masses of the mediastinum. *Radiographics* 2002; **22**(Suppl 1): S79–S93.