Primary pleomorphic liposarcoma of pericardium

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Abstract

We report a case of a 42-year-old man, who presented with a three-weeks’ history of dyspnea and chest oppression induced by light activity, who had a large mass in the pericardium. Computed tomography showed a non-homogeneous density lobulated mass in the pericardial sac. The tumor underwent completely resection and histopathological analysis revealed the tumor to be a pleomorphic liposarcoma, which was composed predominantly of epithelioid cells. The patient is still alive 18 months after the diagnosis was made. The rare incidence of the liposarcoma of the pericardium can easily lead to a misdiagnosis clinically, and the final diagnosis here was made histopathologically.

Keywords: Liposarcoma; Pericardial neoplasms; Thoracic surgery

1. Introduction

Primary cardiac tumors are uncommon; most primary cardiac tumors are also benign. In descending order of frequency (adults) the primary cardiac tumors are: myxomas, fibromas, lipomas, papillary fibroelastomas, rhabdomyomas, and angiosarcomas [1]. The five most common have no malignant potential and account for 80%–90% of all primary heart tumors. Liposarcomas generated from the (intra)pericardium are rarely seen in clinical practice. In fact, liposarcomas are among the most common sarcomas of adult life, which occurred predominantly in the lower limbs and retroperitoneum. This group of tumors can be conceptualized into five main categories on the basis of distinct clinical, pathological and cytogenetical features: atypical lipomatous tumor/well-differentiated liposarcoma, dedifferentiated liposarcoma, myxoid/round cell liposarcoma, pleomorphic liposarcoma and mixed-type liposarcomas. Pleomorphic liposarcoma is the rarest subtype discriminated from other high-grade sarcomas by the presence of pleomorphic lipoblasts. Primary liposarcoma of the pericardium is extremely rare. To the best of our knowledge, there have been no more than nine cases reported in the PubMed database since 1973. Here, we present a case of pleomorphic liposarcoma of the pericardium.

2. Case report

A 42-year-old man presented in October 2008 with a three-weeks’ history of dyspnea and chest oppression induced by light activity. He denied any history of prior cardiovascular and respiratory symptoms before the onset of the present illness. His family history revealed no lipomatous disorders. On examination there was only increased border of cardiac dullness but no signs of pericardial friction rub and cardiac murmur. Laboratory examinations of serum showed higher levels of glutamic oxalacetic transaminase (AST, 88 U/l) and high sensitivity C-reactive protein (hsCRP, 20.96 g/l) but lower levels of albumin (30.99 g/l). Echocardiography showed left atrial enlargement, while right atrial reduction because of the compression of an iso-echoic mass in the pericardium sac with pericardial fluid. The mass was attached to the visceral pericardium and parietal pericardium and did not come into contact with cardiac chambers. Computed tomography (CT) revealed a non-homogeneous density lobulated mass measuring 73 × 109 mm in the pericardial sac, showing calcification, soft tissue signs and fat tissue signs (Fig. 1). The tumor was completely resected, together with a small part of diaphragm and pericardium. The shortest distance between the surgical margin and the tumor edge was about 0.5 cm. Grossly, the tumor was measured as 14 × 10 × 8 cm in size and weighted 750 g. On sectioning, it was yellow, soft and smooth. Histologically, it was composed predominantly of epithelioid cells, which were arranged in large sheets and had round nuclei, often prominent nucleoli, and variably eosinophilic to vacuolated cytoplasm (Fig. 2). Scattered individual or small collections of lipoblasts could be seen. No significant necrosis was present. Immunohistochemical staining showed Vimentin were positive in the cytoplasm of tumor cells (Fig. 2), while S-100, epithelial membrane antigen (EMA), CK7, CK20, CK8/18 and calrectinin were negative in tumor cells (all antibodies from Santa Cruz, Beijing, China).
In April 2009, the patient underwent second surgery because of local recurrence. After that the patient recovered uneventfully, and he is still alive one year after the second surgery, without any radiation and chemotherapy, with no clinical or radiological signs of local recurrence or metastasis.

3. Comments

Liposarcoma usually presents, as in this case, with only compression symptoms, although the tumor is usually large. This may be explained by the slow growth of the tumor mass. Sometimes, the tumor may present with acute pericarditis, cardiac tamponade, multiple organ metastases, syncope and angina [2–4].

As pleomorphic liposarcoma of pericardium is extremely rare, it can easily be misdiagnosed because of the great variety of histological presentations. In fact, the large mass of this case was mistaken as a teratoma after the radiological examination. Even on the frozen section, the epithelioid variant that most resembled a signet-ring cell led us to make a diagnosis of poorly-differentiated carcinoma. Therefore, to make an appropriate histopathological diagnosis, adequate samples, high quality paraffin-embedded, hematoxylin and eosin (HE) stained slides and careful observation are essential. Immunohistochemistry does not play a significant role in recognition of this tumor. The presence of pleomorphic lipoblasts, which is a histological hallmark of pleomorphic liposarcoma, is significant for the recognition of the pleomorphic lipoblasts. In distinguishing the epithelioid subtype from a carcinoma, Vimentin and EMA are helpful markers. Pleomorphic liposarcoma does not express EMA, while carcinoma does not express Vimentin [5]. Compared to EMA, cytokeratins have less value as up to 50% of epithelioid pleomorphic liposarcoma show focal immunoreactivity for cytokeratins [6]. S-100 protein immunoreactivity can be observed in up to 48% of lipogenic areas, while smooth muscle antigen (SMA) was positive in 49% of non-lipogenic areas of pleomorphic liposarcoma [7].

In distinguishing from carcinoma, a panel of immunohistochemical stains is essential. Moreover, we performed calrectinin staining to exclude mesothelioma.

Prognosis of pleomorphic liposarcoma is influenced by some factors: age, site, tumor size, surgical margins, surgical procedure, and radiation therapy. In a preliminary report by Downes et al., the mean time of recurrence after the initial excision was 14 months and the mean time to developing metastases was 19.5 months [5]. Complete tumor resection and adjuvant radiation therapy contribute to reduce the risk of local recurrence and metastasis [8].

We presented a case of a 42-year-old male, with pleomorphic liposarcoma in the pericardium, which have been mistaken for teratoma after radiological examination and carcinoma intraoperation. Our case reveals the importance of histopathological analysis with high quality HE stained slides and immunohistochemical stains.

References

We have read with great interest the article by Wang et al. concerning a case of a primary pericardial pleomorphic liposarcoma [1]. Primary liposarcoma of the mediastinum is extremely rare, representing <1% of mediastinal tumors with <150 cases reported in the literature. It usually occurs in adults, with most cases occurring in patients over 40 years old [2]. Liposarcoma develops more commonly in the posterior mediastinum. Liposarcoma of the anterior mediastinum is very rare and only a few cases have been reported in the international literature [3].

We have treated a similar patient three years ago, with good results concerning recurrence and survival till now [4]. We would like to highlight that recurrence is common in deep-seated liposarcomas and that it becomes apparent within the first six months in most cases, but it may be delayed for five or 10 years following the initial excision. Recurrence is related mainly to the incomplete excision and tumor tissue left behind at the time of surgery [5]. Therefore, a close follow-up is strongly recommended.

References


eComment: Primary pleomorphic liposarcoma: a rare mediastinal tumor

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