

Incidental Finding Of a Massive Cardiac Metastasis in a Boy Treated For a Parameningeal Rhabdomyosarcoma

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Abstract

An eight-year-old boy treated for a local recurrence of a stage IV parameningeal rhabdomyosarcoma complains four months after a second complete response, from dyspnea, chest pain and cyanosis. Imagery showed a bulky pleuro-mediastinal lesion. Echocardiography revealed a massive right atrial mass suggestive of metastasis. A partial response (70%) was obtained after four courses of chemotherapy, but progression was observed after ten cycles of chemotherapy and patient died one month later.

This case of incidentally diagnosed heart metastases is particular because it occurred in a child and the primary tumor was a parameningeal rhabdomyosarcoma.

Keywords: Rhabdomyosarcoma; Heart Neoplasms; Neoplasm Metastasis

Introduction

In oncology, sarcomas are responsible for 15% of metastatic disease, predominantly to lungs. Cardiac metastases are extremely rare especially in children but their incidence is probably under-estimated. Those arising from sarcomas are exceptional [1]. They are usually late and clinically silent. Treatment of cardiac metastases is controversial because of its rarity. We report one exceptional case of metachronous cardiac metastasis from a pediatric rhabdomyosarcoma, focusing on diagnostic and therapeutic difficulties.

Case report

An eight-year-old boy with no relevant previous medical history was treated in our institute for an embryonal rhabdomyosarcoma of the infratemporal fossa. Lung metastases with balloon release were diagnosed but there was no mediastinal involvement.

A complete remission was obtained after 4 courses of IVADO (ifosfamide, doxorubicin, mesnum, actinomycin and vincristin) and 5 subsequent courses of IVA (ifosfamide, mesna, actinomycin and vincristin). Treatment was completed with local radiation

therapy at the total dose of 54 Gy. Two months after complete response, the patient was readmitted with dyspnea, chest pain and cyanosis. There were no cardiac failure symptoms. Chest CT scan did not detect pulmonary metastases but showed multiple pleural and mediastinal nodules with a heart and mediastinal deviation (Figure 1). Pleural biopsy confirmed the relapse.

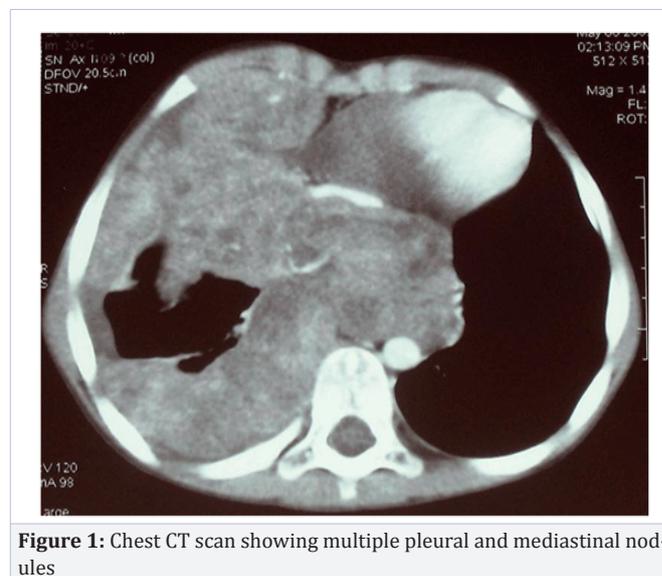


Figure 1: Chest CT scan showing multiple pleural and mediastinal nodules

Subsequent transthoracic echocardiogram performed in order to evaluate the left ventricular ejection fraction before initiating salvage chemotherapy revealed a suspect massive right atrial metastasis (Figure 2). Salvage chemotherapy by ifosfamide and vinorelbine leads to a partial response, evaluated at 70% after four courses. Disease progression was diagnosed after ten courses of chemotherapy and patient died one month later.



Figure 2: Echocardiogram showing a massive right atrial metastasis

Discussion

The most common tumors with cardiac metastatic potential are carcinomas and melanomas [1] which are rare tumors in children. Very few papers concerned cardiac metastases from pediatric cancers [2].

Neoplasms can metastasize to the heart by one of four different pathways: lymphatic, hematogenous, direct extension and transvenous extension [3]. If we consider these mechanisms, secondary heart tumors would not be uncommon.

But are cardiac metastases so rare? The frequency of cardiac metastases is generally underestimated since they were found in about 25% of postmortem patients (children and adults) who had died of malignancies [1, 4].

Previous reports have emphasized the difficulty in the premortem detection of cardiac metastases [4] in part because only 15% of patients are symptomatic. Cardiac metastases misdiagnosis could be due to the fact that they are usually late and clinically silent. Moreover, as in our patient, thoracic symptoms are often attributed to pulmonary metastases which are the most frequent metastasis site in sarcomas. To our best knowledge, there are 20 cases of cardiac metastases of soft tissue sarcoma reported in the literature [3, 5-11], all of them were found in adult patients. Our case is the first case reported of cardiac metastases of a soft tissue sarcoma in a child. In our patient, dyspnea, tachypnea and chest pain were thought to result from the pleuro-mediastinal metastases. Cardiac lesion was not suspected even on the chest CT-scan.

Antemortem diagnosis of cardiac metastases is really important, since it is immediately connected with the discussion of operability, as well as with the choice of the most adapted surgical or medical therapeutic approach [3].

According to K. Reynen [1], heart sarcomas have to be considered metastatic if an extracardiac tumor site has already been revealed by clinical examination, by diagnostic procedures, or post-mortem i.e cardiac metastases mostly appear in patients with disseminated tumor disease and heart solitary metastases are very rare.

In contrast to primary sarcomas, metastatic sarcomas involve both the right and left ventricles with varying degrees of extension into the epicardium or pericardium [6]. Transthoracic and transesophageal echocardiography are the most widely used diagnostic modalities. Magnetic resonance imaging (MRI) may be helpful for selected cases [8]. Endocardial biopsy or thoracotomy, whenever possible, is recommended in order to have a pathological diagnosis [2].

Treatment of cardiac metastases is controversial because of its rarity. Chemotherapy and/or radiotherapy are the most commonly proposed treatment modalities. Surgery is indicated when there is a hemodynamic decompensation [3].

Conclusion

This original report illustrates that cardiac metastases should be considered in the differential diagnosis for a thoracic symptomatology in patients treated for head and neck soft tissue sarcoma since symptoms are not specific and routine investigations (CT-scan) are not accurate. Echocardiography and MRI are more likely to be contributive.

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