https://doi.org/10.30702/ujcvs/23.31(04)/ChB050-126129
UDC 616.718.4-006.33-033.1:616.24-005]-089

Amulya Cherukumudi, Resident, https://orcid.org/0000-0001-5067-1126
Bhagyashree R., Assistant Professor, https://orcid.org/0000-0001-7950-4575
Ramesh C. Hebbale, Associate Professor, https://orcid.org/0000-0003-4988-7346
Varadaraju R., Professor, https://orcid.org/0000-0002-6140-7395
Department of Cardiovascular and Thoracic Surgery, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bangalore, India

A Case of Hip Chondrosarcoma Metastasis to the Right Ventricle

Abstract

Background. While primary cardiac tumors occur rarely, it is observed that cardiac metastases are almost 20 times more common. These are, however, observed during autopsy and seldom missed in living patients. Patients with cardiac metastases present commonly with dyspnea on exertion or pleuritic chest pain. Most common site of intracardiac metastases is right atrium.

Case presentation. Here we present a case of a 21-year-old male who presented for a routine check-up to the cardiac outpatient department for assessment of fitness for chemotherapy. He was a known case of right hip chondrosarcoma for 2 years, being treated with Ayurvedic medications. He had no symptoms related to the respiratory or cardiac system. He was afebrile during the examination, however, was restricted to a bed due to the pain in his right hip. On evaluation, the patient was diagnosed with a right ventricular thrombus extending to the pulmonary artery (PA) causing an impending pulmonary embolism. On opening the right atrium and PA, we found a loose, whitish-grey mass, adherent to the right ventricular papillary muscle, entangling the chordae. The tricuspid valve leaflets were also found to be tethered to this mass. The mass was found to extend to the PA beyond the leaflets, however, the leaflets were free of the tissue. The mass was extracted piece-meal through right atrium and PA approach. Histopathology revealed metastatic chondrosarcoma tissue.

Conclusion. Cardiac metastases of chondrosarcoma are a rare condition, and can be often asymptomatic. Such patients often present with dyspnea, and imaging may not be able to rightly identify the cause. A high index of suspicion is necessary in patients with a known current or prior malignancy before committing to surgical intervention.

Keywords*: bone tumors, carcinoma, metastatic complications, dyspnea, hip pain, case report.*

Introduction. Cardiac metastases are nearly 20 times more frequent and have been identified in autopsy series in up to one in five cancer patients [1]. Primary cardiac tumors, however, are very rare. Moreover, cardiac metastases of chondrosarcoma are quite uncommon. There are only few available case reports of such an occurrence; to our knowledge, this is the fourth report and the second such report secondary to right hip chondrosarcoma [1, 2].

A malignant tumor of cartilaginous tissues, chondrosarcoma, hardly ever develops in the heart. It is assumed that it develops from multipotent mesenchymal stem cells that malignantly differentiate into cartilage [3, 4].

Most patients are symptomatic, presenting with features of acute onset dyspnea, pleuritic chest pain or features of congestive heart failure. These cases often progress rapidly, and hence are detected during an autopsy. It is rare to find a perfectly asymptomatic patients with extensive metastases.

It is suggested in literature that cardiac metastases are most frequently noted in right atrium, due to the venous drainage being received from the systemic circulation. It indicates poor prognosis in such patients, with extensive spread.

We present this unique, rare case of right ventricular (RV) metastasis of chondrosarcoma originating from the right iliac bone.

Case report. This is a case of a 21-year-old male who complained of insidious onset of right hip pain and restriction of movement over the last three years. Two years back, he was diagnosed with right hip chondrosarcoma. He underwent full evaluation at that point, and was advised chemoradiotherapy followed by surgery. However, they decided to take alternative (Ayurvedic) medicine instead

of chemoradiotherapy and surgery. When his right hip pain aggravated, they sought advise at hospital, wherefrom he was referred for assessment of fitness for chemotherapy. He had no symptoms related to the respiratory or cardiac system. There was no family history of malignancy or heart disease. He did not smoke, consume alcohol or chew tobacco. He had not undergone any surgery in the past.

Positron emission tomography (PET) was done prior to referral, which showed disease limited to the right ilium (Fig. 1).

He was afebrile during the examination, however, was restricted to a bed due to the pain in his right hip. There was localized tenderness and rise of temperature in the right hip. Cardiovascular, respiratory and abdominal examination was found to be grossly normal.

Blood parameters revealed anemia with leukocytosis, but the rest of the findings were normal.

2D echocardiography revealed an RV thrombus extending to the pulmonary artery (PA) causing an impending pulmonary embolism. The patient was referred for emergency surgery.

The approach followed was a midline sternotomy, vertical pericardiotomy and aorto-bicaval cannulation. Cardiopulmonary bypass was established and the patient was cooled to 30 degrees centigrade. On external inspection of the heart and pericardium, no gross abnormality was noted. After right atriotomy, we found a loose, whitish-grey mass, adherent to the RV papillary muscle, entangling the chordae (Fig. 2, 3).

The tricuspid valve leaflets were also found to be tethered to this mass. However, it was decided to leave the leaflets as they are.

The mass was found to extend to the PA beyond the leaflets, however, the leaflets were free of the tissue. The mass was extracted piece-meal through right atrium (RA) and PA approach. (Both approaches were used to extract the tumor, from tricuspid valve to pulmonary valve.) In the immediate postoperative period, the patient had serous discharge from the wound and repeated histopathology revealed metastatic chondrosarcoma tissue (Fig. 4). Based on this report, the patient was referred for chemotherapy +/– radiotherapy. However, the patient developed fever on postoperative day 8, and succumbed to the disease in the hospital we referred him to.

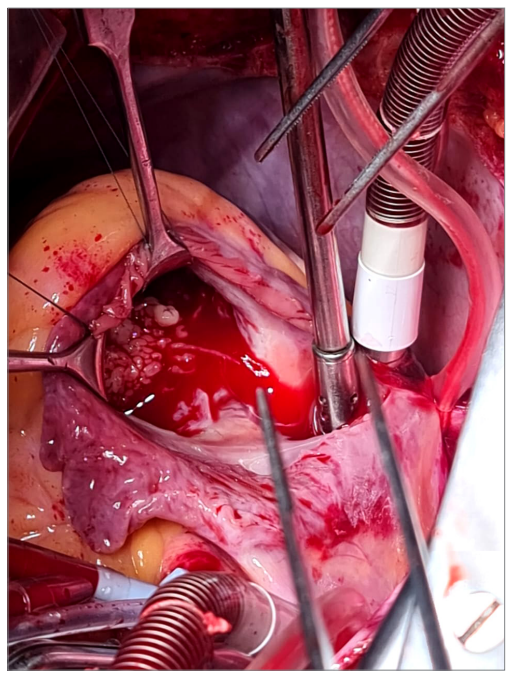

Fig. 2. *Large, irregular loose mass, whitish-grey, adherent to papillary muscles and chordae*

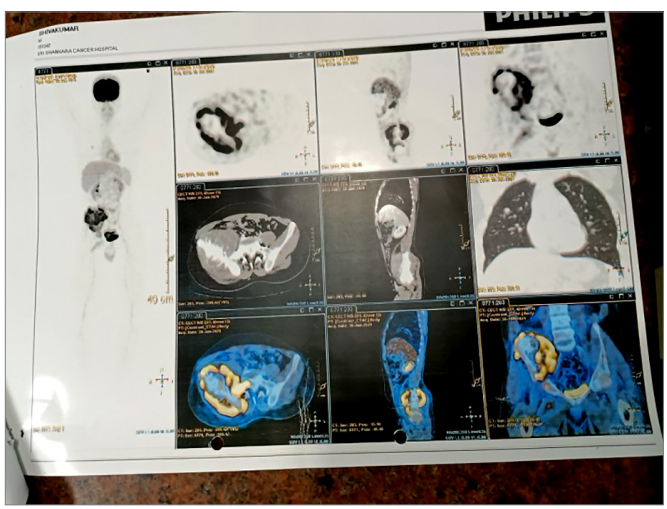

Fig. 1. *PET scan showing metabolically active lesion in the right iliac bone with localized swelling and inflammation. There is no evidence of metastases in the cardiac*

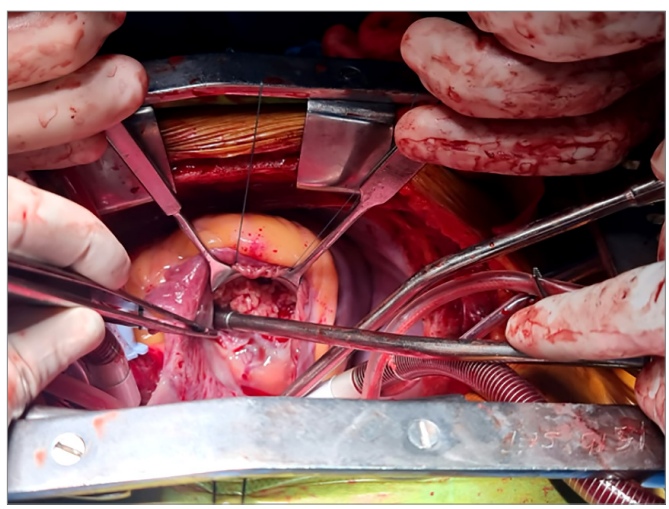

Fig. 3. *Loose mass tethered to tricuspid valve leaflets*

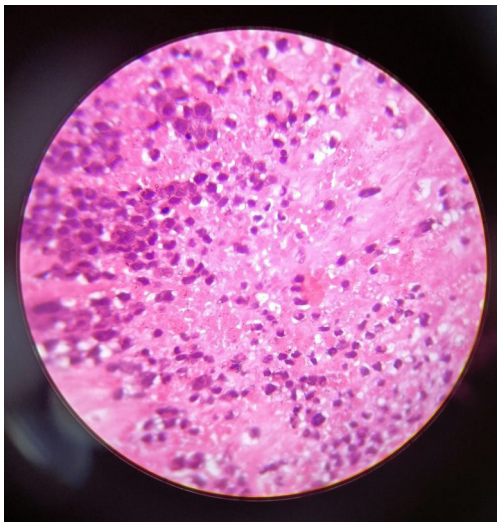

Fig. 4. *Histopathology (40x magnification) showing sheets of mesenchymal cells with increased nucleo-cytoplasmic ratio suggestive of metastases*

Discussion. Cardiac metastases are uncommon, with RA being the most common site. It is rare for the RV to be involved sparing the RA [1]. Chondrosarcoma of the bone is known to be rare, but it is still the third most common bone tumor [2]. It is locally aggressive in most of its forms, though metastases are rare. Moreover, this is the first case where the patient was asymptomatic and the imaging did not provide the diagnosis.

We need to keep in mind that patients with primary malignancy in any part of the body may develop cardiac metastases. Even the most specific imaging modality may not differentiate between tumor and thrombus, creating an unusual dilemma for surgeons [4]. In the case report presented by Haslinger et al, they faced a similar challenging situation, where patient was suspected to have pulmonary embolism which later intraoperatively was identified as metastases [4]. They suggested that the peripheral rim enhancement is due to the slowed blood flow within the chambers.

In a study by Leung et al [5], progression of primary chondrosarcoma to cardiac metastases took about 2-3 years, with median survival of those with cardiac involvement being 2 months. Other than providing debulking surgery, little can be offered for such patients with extensive involvement of the disease. Adequate pre-operative planning and treatment is necessary to prevent deposits of such tissue and disseminating the tumor tissue through the bloodstream. That being said, this is a life-saving procedure in cases with impending pulmonary embolism [6].

Conclusion. Cardiac metastases of chondrosarcoma are extremely rare, and represent a very late presentation and poor prognosis. Such patients often present with dyspnea, and imaging may not be able to rightly identify the cause. A high index of suspicion is necessary in patients with a known current or prior malignancy before committing to surgical intervention.

References

1. Maurea N, Ragone G, Coppola C, Caronna A, Tocchetti CG, Agozzino L, et al. Intracardiac metastasis originated from chondrosarcoma. J Cardiovasc Med (Hagerstown). 2017;18(5):385-388. https://doi.org/10.2459/JCM.0b013e32834165eb
2. Gelderblom H, Hogendoorn PC, Dijkstra SD, van Rijswijk CS, Krol AD, Taminiau AH, et al. The Clinical Approach Towards Chondrosarcoma. Oncologist. 2008;13(3):320-329. https://doi.org/10.1634/theoncologist.2007-0237
3. Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumours: diagnosis and management. Lancet Oncol. 2005;6(4):219-228. https://doi.org/10.1016/S1470-2045(05)70093-0
4. Haslinger M, Dinges C, Granitz M, Klieser E, Hoppe UC, Lichtenauer M. Right Heart Failure Due to Secondary Chondrosarcoma in the Right Atrium. Circ Cardiovasc Imaging. 2020 Apr;13(4):e009824. https://doi.org/10.1161/CIRCIMAGING.119.009824
5. Leung CY, Cummings RG, Reimer KA, Lowe JE. Chondrosarcoma Metastatic to the Heart. Ann Thorac Surg. 1988;45(3):291-295. https://doi.org/10.1016/s0003-4975(10)62465-4
6. Bansal D, Gupta A, Bhattacharya S, Kumar S, Gupta VK, Aiyer P. Recurrent high-grade chondrosarcoma abutting the right ventricular outflow tract. Asian Cardiovasc Thorac Ann. 2019;27(6):501-504. https://doi.org/10.1177/0218492319840967

Випадок метастазування хондросаркоми стегнової кістки у правий шлуночок серця

Амулья Черукумуді (Amulya Cherukumudi), ординатор
Бхагьяшрі Р. (Bhagyashree R.), доцент
Рамеш Ч. Хеббале (Ramesh C. Hebbale), доцент
Варадараджу Р. (Varadaraju R.), професор

Інститут серцево-судинних наук та досліджень імені Шрі Джайадева, Бангалор, Індія

Резюме. Первинні пухлини серця виникають рідко, однак метастази в серце розвиваються майже у 20 разів частіше. Їх виявляють під час аутопсії, і зрідка вони залишаються недіагностованими у живих пацієнтів. У пацієнтів із метастазами в серце часто спостерігають задишку під час фізичного навантаження з плевритичним болем у грудній клітці. Найчастіше метастази локалізуються у правому передсерді.

Опис випадку. Чоловік віком 21 рік звернувся для планового огляду до амбулаторного кардіологічного відділення для визначення можливості проведення хіміотерапії. Він мав підтверджену хондросаркому правого стегна протягом 2 років і отримував лікування аюрведичними засобами. Симптоми з боку дихальної або серцево-судинної системи були відсутні. Під час обстеження він мав нормальну температуру, однак йому було призначено постільний режим через біль у правому стегні. Шляхом обстеження у пацієнта виявлено тромб у правому шлуночку з поширенням у легеневу артерію, що спричинило загрозу тромбоемболії легеневої артерії. Після розтину правого передсердя і легеневої артерії ми виявили пухке біло-сіре новоутворення, що прилягало до папілярного м'яза правого шлуночка, залучивши хорду. Стулки тристулкового клапана також були прикріплені до цього новоутворення. Новоутворення поширювалося до легеневої артерії за стулками, однак стулки його тканинами уражені не були. Новоутворення було видалено частинами через праве передсердя і легеневу артерію. Гістопатологічне дослідження виявило тканини метастатичної хондросаркоми.

Висновки. Метастази хондросаркоми в серці – рідкісне захворювання, що часто може бути безсимптомним. У таких пацієнтів часто спостерігається задишка, і візуалізація не завжди дозволяє правильно визначити причину. Пацієнти з відомим наявним або попереднім злоякісним новоутворенням потребують високого рівня підозри до проведення хірургічного втручання.

Ключові слова: *пухлина кістки, карцинома, метастатичні ускладнення, задишка, біль у стегні, опис випадку.*

Стаття надійшла в редакцію / Received: 03.10.2023
Після доопрацювання / Revised: 20.10.2023
Прийнято до друку / Accepted: 18.12.2023