ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 9 | September - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PRIMARY CARDIAC LEIOMYOSARCOMA: A RARE CASE REPORT

Pathology

Dr. Anchana Gulati Associate Professor, Department of Pathology, IGMC Shimla, India- 171001.

Dr. Deepika Sharma* Junior Resident, Department of Pathology, IGMC Shimla, India- 171001. *Corresponding Author

Dr. Sudarshan Kumar Sharma Professor & Head Department of Pathology, IGMC Shimla, India- 171001.

Dr. Sudhir Mehta Professor & Head Department of CTVS, IGMC Shimla, India- 171001.

ABSTRACT

Primary cardiac leiomyosarcoma is a rare entity comprising less than 1 % of all primary cardiac malignancies. We present a case of primary left atrial leiomyosarcoma in a 60 years old female patient who presented with dyspnoea. It was preoperatively diagnosed as atrial myxoma. On careful histopathological examination it was diagnosed as cardiac leiomyosarcoma and confirmed on immunohistochemistry.

KEYWORDS

leiomyosarcoma, atrial myxoma.

INTRODUCTION

Primary cardiac tumors are rare with a prevalence of 0.001% to 0.28%. Also majority are benign, only 25% are malignant[1]. Metastatic cardiac tumors are 100- fold more common than primary lesions. Mostly, primary cardiac sarcomas are confused with myxoma which is the most common benign tumor. Among primary malignant cardiac tumors, most common are undifferentiated ones followed by angiosarcomas, rhabdomyosarcoma and fibrosarcoma[2]. Leiomyosarcoma represents less than 1% of primary cardiac malignancies. We present a case aimed to highlight clinical and histopathological features of primary cardiac leiomyosarcoma.

Case presentation

A 60 years old female patient presented to cardiology department with chief complaints of swelling bilateral extremities since 9 months and shortness of breath for the last 3 months.

On general physical examination there was peripheral edema only.
On contrast enhanced computerized tomography scan of the thorax: there was a large hypodense lesion of size 54x48 mm with patchy areas of mild contrast enhancement noted in mediastinum possibly in left atrium.

On echocardiography: there was a large left atrial mass causing significant mitral valve inflow obstruction. Possibility of atrial myxoma was given.

Surgical excision of tumor was uneventful and specimen sent to pathology department. We received a single well circumscribed soft tissue mass of size 5x4x1.5cm for histopathological examination. Grossly it was irregular gray brown and on cut section revealed gray white, gray brown and yellowish solid areas.

Microscopic examination revealed tumor cells arranged in fascicular pattern with focal areas of necrosis and myxoid change. Tumor cells were spindle shaped having moderate pleomorphism, cigar shaped vesicular nuclei with blunt end, intranuclear cytoplasmic inclusions, variably prominent nucleoli and eosinophilic fibrillary cytoplasm. Mitotic figures were 5 per 10 high power fields.

Immunohistochemically these tumor cells showed positivity for vimentin, smooth muscle actin and were negative for cytokeratin and S-100. Ki 67 index was 10 %. These findings confirmed the diagnosis of primary leiomyosarcoma of left atrium.

DISCUSSION

Cardiac tumors are rare and majority of these are metastatic lesions with primary usually in the breast, lung or of renal origin. Primary tumors of the heart are mostly benign. Leiomyosarcoma is a malignant mesenchymal tumor with smooth muscle differentiation and comprised 10 % of primary cardiac sarcomas[3]. It most commonly occurs in fourth decade with slight female predilection[4]. It has two usual sites of origin. One is the left atrium where it may present as single or multinodular endocavitary mass and mimics left atrial myxoma,. Second is the pulmonary infundibulum where it may present abruptly mimicking pulmonary embolism. Leiomyosarcoma is believed to arise from pulmonary smooth muscle cells[5]. They may also arise in right ventricle, right atrium and left ventricle in about 20.2 per cent, 15.2 per cent and 5.1 per cent cases respectively[6].

The patient usually remains asymptomatic until advanced stages. Clinical presentation is atypical, however most patients presents with dyspnea, cough, chest pain and edema due to cardiac outlet obstruction[7]. Some patients have palpitations and tachycardia associated with arrhythmia due to myocardial involvement. Non-specific symptoms like fatigue, fever, malaise delay the diagnosis. Transthoracic 2-D echocardiographic imaging is the most sensitive imaging technique to diagnose cardiac tumors with a sensitivity of nearly 100 per cent. It can detect tumor as small as 3 mm. Soft-tissue characterization remains almost similar as with CT scan and cardiac MRI but is outlined by cardiac MRI. Myocardial involvement such as tumor infiltration not clearly depicted by echocardiography. Tumor has to be around 1 cm in size before becoming detectable by MRI and CECT scan. Grossly they present as friable solid sessile, intraluminal mass. In 30% cases they may present as multiple nodules[8]. Histopathologically they appears as compact fascicles of spindle cells with blunt ended nuclei and eosinophilic cytoplasm containing glycogen and peri-nuclear vacuoles. There are areas of necrosis and increased mitotic activity. Immunohistochemically these lesions are positive for smooth muscle actin and desmin. Cardiac leiomyosarcoma has a poor prognosis due to advanced stage at presentation. Complete surgical resection is the mainstay of treatment followed by chemotherapy with doxorubicin, ifosfamide and dacarbazine recommended for use[9]. Radiotherapy is usually a palliative therapy.

Cardiac leiomyosarcoma has a high rate of local recurrences and metastases even after optimal resection of the tumor. On review of the reported cases of cardiac leiomyosarcomas, the mean survival rate of patients who underwent surgery and chemotherapy was about 12 months. Several factors associated with worse prognosis include high tumor grade, high mitotic index, positive surgical margins and metastasis[10]. Early histopathological diagnosis is important for choice of treatment and better prognosis. Our patient did not have any detectable metastasis at the time of diagnosis.

CONCLUSION

Primary cardiac malignancies are rare and clinical distinction from the more common atrial myxoma is a diagnostic problem. Hence histopathologic evaluation followed by immunohistochemistry are the mainstay of diagnosis. Treatment is a combination of surgery and the chemotherapy but the prognosis is dismal.

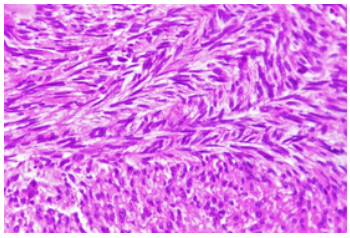

Figure 1: fascicular arrangement of tumor cells (400X).

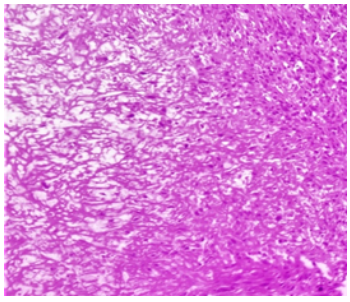

Figure 2: tumors cells with myxoid and necrotic areas (100X).

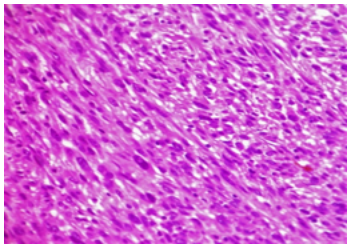

Figure 3: tumor cells with cigar shaped blunt ended nuclei and intranuclear inclusions (400X).

REFERENCES

1. Talukder M, Joyce L, Marks R, Kaplan K. Primary cardiac synovial sarcoma. Interactive CardioVascular and Thoracic Surgery. 2010 Oct 1;11(4):490-2.
2. Andersen RE, Kristensen BW, Gill S. Cardiac leiomyosarcoma, a case report. International Journal of Clinical and Experimental Pathology. 2013;6(6):1197.
3. Travis LB, Ng AK, Allan JM, Pui CH, Kennedy AR, Xu XG, Purdy JA, Applegate K, Yahalom J, Constine LS, Gilbert ES. Second malignant neoplasms and cardiovascular disease following radiotherapy. Journal of the National Cancer Institute. 2012 Mar 7;104(5):357-70.
4. Wang JG, Cui L, Jiang T, Li YJ, Wei ZM. Primary cardiac leiomyosarcoma: an analysis of clinical characteristics and outcome patterns. Asian Cardiovascular and Thoracic Annals. 2015 Jun;23(5):623-30.
5. Parissis H, Akbar MT, Young V. Primary leiomyosarcoma of the right atrium: a case report and literature update. Journal of cardiothoracic surgery. 2010 Dec;5(1):1-4.
6. Uy Crizel Denise V. Primary Leiomyosarcoma of the Left Atrium. Case Reports: Open Access. 2019 Oct;4:1-4.
7. Behi K, Ayadi M, Mezni E, Meddeb K, Mokrani A, Yahyaoui Y, Ksontini F, Rais H, Chrait N, Mezlini A. Two years survival of primary cardiac leiomyosarcoma managed by surgical and adjuvant therapy. Clinical sarcoma research. 2017 Dec 1;7(1):5.
8. Araoz PA, Eklund HE, Welch TJ, Breen JF. CT and MR imaging of primary cardiac malignancies. Radiographics. 1999 Nov;19(6):1421-34.
9. Bakaeen FG, Jaroszewski DE, Rice DC, Walsh GL, Vaporciyan AA, Swisher SS, Benjamin R, Blackmon S, Reardon MJ. Outcomes after surgical resection of cardiac sarcoma in the multimodality treatment era. The Journal of thoracic and cardiovascular surgery. 2009 Jun 1;137(6):1454-60.
10. Lv Y, Pang X, Zhang Q, Jia D. Cardial leiomyosarcoma with multiple lesions involved: a case report. Int J Clin Exp Pathol. 2015;8(11):15412–6.