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Case report / Olgu Sunumu

Cardiac metastasis of clear cell sarcoma

Berrak hücreli sarkomun kardiyak metastazı

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Clear cell sarcoma is a rare soft tissue tumor. Cardiac metastasis of clear cell sarcoma is seen even rarely. A 34-year-old female patient was admitted to our hospital with the complaints of pain and swelling on the posterior aspect of her right knee. She was operated and the popliteal mass was removed with clean surgical borders. Fourteen months following diagnosis, the patient applied to our clinic with a metastatic mass of 47x33 mm size at the junction of the right atrium and right ventricle. A pericardial window was opened and pericardial resection was made through left anterolateral thoracotomy. Biopsy of the lesions in the right ventricle showed similar findings with the popliteal tumor and was reported as clear cell carcinoma metastasis. The patient was re-hospitalized in the second month of the follow-up. Cardiac arrest occurred during the supportive therapy. Emergent median sternotomy was performed. Tumor mass had grown extremely large. The patient died. In this article, metastasis of clear cell sarcoma to the heart was reported for the first time. The involvement of heart by clear cell sarcoma was aggressive. Although pericardial resection and opening a pericardial window provided temporary relief, the prognosis was fatal due to rapid growth of the tumor.

Key words: Cardiac metastasis; clear cell sarcoma; soft tissue tumors.

Berrak hücreli sarkom, nadir görülen bir yumuşak doku tümörüdür. Berrak hücreli sarkomun kardiyak metastazı daha da seyrek görülür. Otuz dört yaşında kadın hasta hastanemize sağ dizinin arka bölümünde ağrı ve şişlik yakınmasıyla başvurdu. Ameliyat edilen hastadaki popliteal kitle cerrahi sınırları temiz olarak çıkarıldı. Hasta tanıdan 14 ay sonra kliniğimize sağ atriyum-sağ ventrikül bileşkesinde 47x33 mm boyutlarında metastatik kitle ile yeniden başvurdu. Perikardiyal pencere açılarak, sol anterolateral torakotomi ile perikardiyal rezeksiyon yapıldı. Sağ ventrikküldeki lezyonun biyopsisi primer popliteal tümörünki ile benzer yapıda berrak hücreli karsinoma metastazı ile uyumlu olarak bildirildi. İzleme alınan hasta iki ay sonra yeniden hastaneye yatırıldı. Hasta destek tedavisi sırasında kardiyak arreste girdi. Acilen median sternotomi yapıldı. Tümör kitlesi çok büyümüştü. Hasta kaybedildi. Bu yazıda berrak hücreli sarkomun kalbe metastazı ilk kez bildirilmektedir. Berrak hücreli sarkom kalpte agresif yayılım göstermişdi. Yapılan perikard rezeksiyonu ve bir perikardiyal pencere açılması geçici rahatlama sağladıysa da tümörün hızlı büyümesi nedeniyle ölümcül seyretti.

Anahtar sözcükler: Kardiyak metastaz; şeffaf hücreli sarkom; yumuşak doku tümörleri.

Clear cell sarcoma is a rare soft tissue tumor that was first defined by Enzinger in 1965.[1] It constitutes approximately 1% of all soft tissue sarcomas and usually occurs in the lower extremities of young adults in the form of a slow-growing soft tissue mass presenting with pain. The prognosis is discouraging as it tends to metastasize to distant organs. The aim of this report is to share our experience involving a case of rare cardiac metastasis of clear cell sarcoma with the medical community since a search of the literature produced no similar reports.

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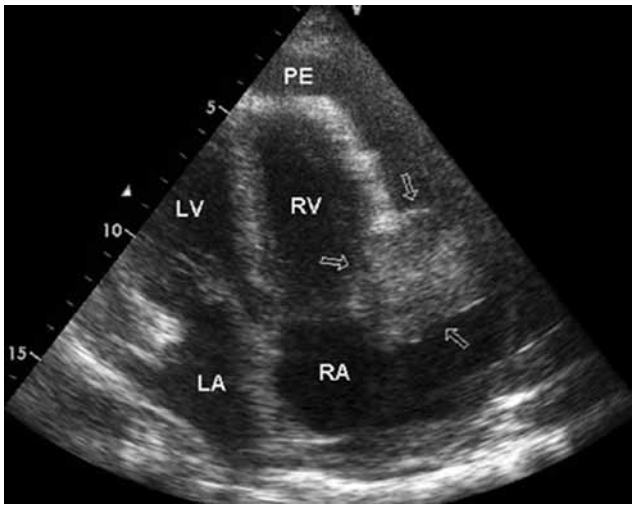

Figure 1. Echocardiographic examination showing the metastatic mass (arrows indicate the mass). PE: Pulmonary embolism; LV: Left ventricle; RV: Right ventricle; LA: Left atrium; RA: Right atrium.

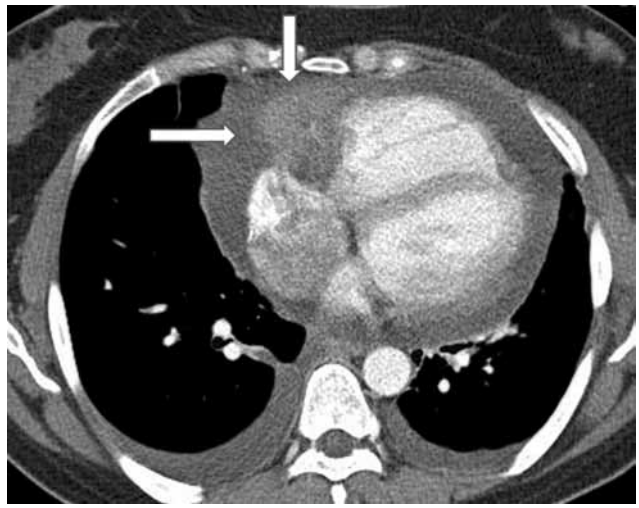

Figure 2. Computed tomography revealing a mass of 49x50 cm adjoining the right atrium and right ventricle that is indicative of metastasis (arrows show the mass).

CASE REPORT

A 34-year-old female patient was admitted to our hospital complaining of pain and swelling on the posterior aspect of her right knee. Magnetic resonance imaging (MRI) confirmed a soft tissue mass (38x58x68 mm) in her right popliteal fossa, and surgery was planned for tissue diagnosis and treatment. A wide excision of the mass was performed, and a pathological diagnosis revealed clear cell sarcoma with clean surgical borders. Advanced examinations done at the time of the operation showed no metastasis, and local adjuvant radiotherapy and chemotherapy (doxorubicin, ifosfamide, and mesna) were administered.

Fourteen months later, the patient was admitted to the hospital emergency room with complaints of hypotension, palpitation, and dyspnea. In her echocardiographic examination, pericardial effusion causing cardiac compression and a mass of 47x33 mm size at the junction of the right atrium and right ventricle were detected (Figure 1). The effusion was drained by pericardiocentesis. A cytological study of the pericardial fluid revealed findings suggestive of clear cell sarcoma metastasis. Computed tomography (CT) revealed a mass of 49x50 cm size adjoining the right atrium and right ventricle indicative of metastasis (Figure 2). Further surgical treatment was recommended to the patient, but she declined intervention.

Pericardiocentesis was performed four times due to recurrent pericardial effusion. Thereupon, the patient became convinced of the need for surgery to open a pericardial window and carry out pericardial resection to the extent that the mass permitted. She was operated on via a left anterolateral thoracotomy, the pericardial window was opened, and the pericardial resection was made (Figure 3). There was tumor invasion in the lateral right ventricle segments and the epicardium. She was discharged on postoperative day four after an uneventful recovery.

Pathological examination

A light microscopic evaluation of the resected popliteal tumor revealed tumor nests composed of uniform cells with clear to eosinophilic cytoplasms and round-oval

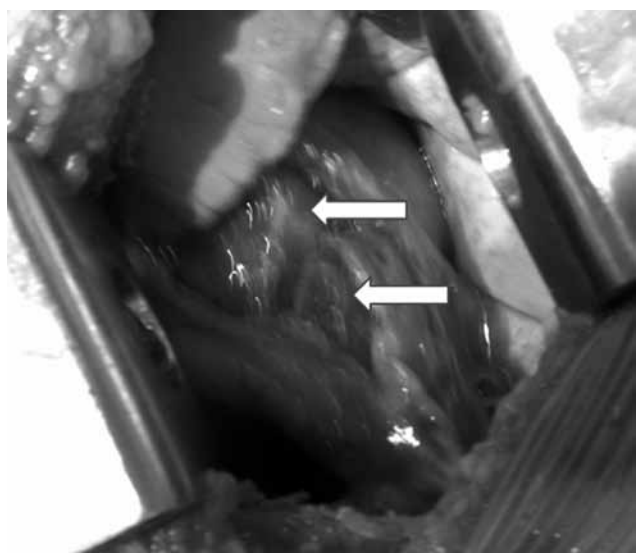

Figure 3. Photograph of the cardiac tumor in the first operation (anterolateral thoracotomy) (arrows show tumor).

nuclei containing central nucleoli (Figure 4a). Foci of necrosis were also observed. An immunohistochemical study was positive for S-100 protein, human melanoma black-45 (HMB-45) (Figure 4b), bcl-2, vimentin, and epithelial membrane antigen (EMA), but it was negative for CD68, alpha-smooth muscle actin (SMA), desmin, myogenin D, CD 99, and CD 34. The combined data led to diagnosis of the tumor as clear cell sarcoma.

The macroscopic appearence of pericardial fluid was hemorrhagic. Multiple cytospin slides stained with hematoxylin-eosine (HE) and Papanicolau stains and cell blocks were prepared. In a microscopic evaluation, numerous macrophages, small groups of mesothelial cells, and scattered round-to-oval cells resembling macrophages containing eccentrically located nuclei and single nucleoli were observed, Some of these cells were binuclear (Figure 4b) and positive for HMB-45 and vimentin. However, they tested negative for calretinin, Hector Battifora mesothelial epitope-1 (HBME-1), and CD68 in the immunohistochemical study that was performed on the sections taken from the cell block (Figure 4c). The effusion material was reported as "suspicious for malignancy". Furthermore, a biopsy of the lesions in the right ventricle and the biopsy performed on the popliteal tumor, which had indicated clear cell carcinoma metastasis, showed similar findings (Figure 4d).

A control cardiac MRI performed one month after the pericardial resection disclosed that the tumor had spread to both ventricle walls (Figure 5). The patient was hospitalized again in the second month of follow-up with complaints of respiratory distress and hypotension. Cardiac arrest developed during hospitalization, and an emergency median sternotomy was performed. Enormous enlargement of the tumor was observed, and a biopsy was taken from the cardiac mass. Unfortunately, however, the patient did not survive the operation.

DISCUSSION

Clear cell sarcoma appears in the form of a painful soft tissue mass which is characteristically slow-growing. Although it may appear at any age in any part of the body, it often arises in the lower extremities of young adults.[2] In our case, the tumor arose at the back of the

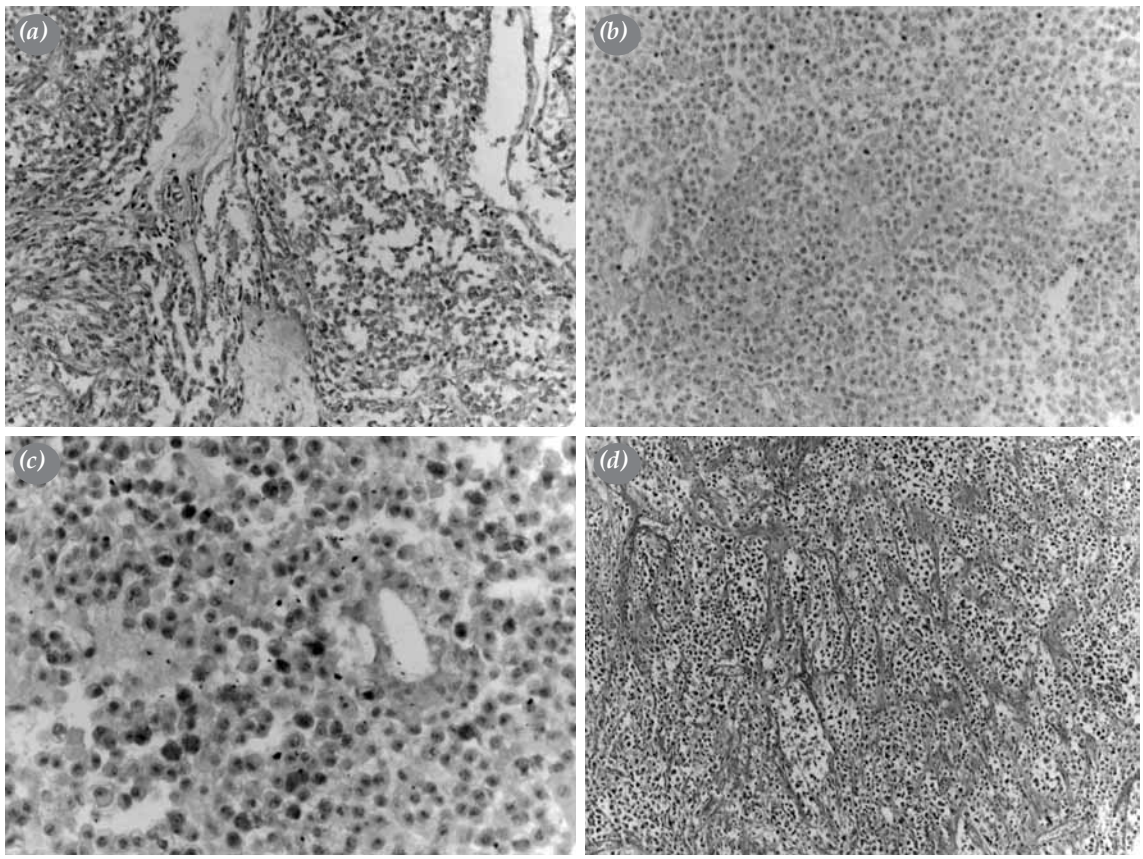

Figure 4. Pathological specimens. **(a)** The primary tumor (H-E x 100). **(b)** Microphotograph of the cell block from effusion (H-E x 100). **(c)** Cytoplasmic HMB-45 positivity in an immunohistochemical study (HMB-45 x 200). **(d)** Tumor cells in the right ventricle biopsies (H-E x 200).

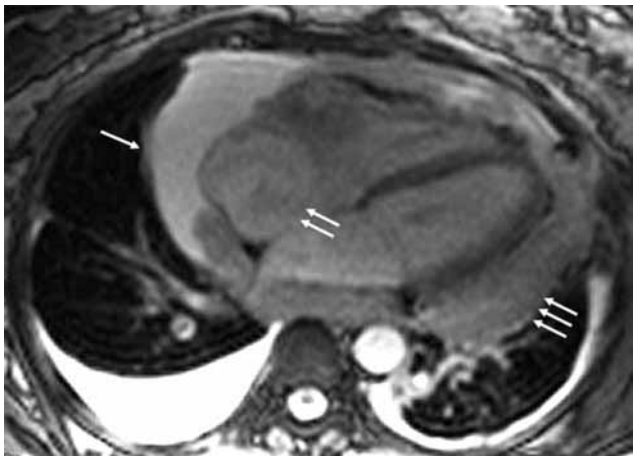

Figure 5. A control cardiac magnetic resonance imaging performed one month after the pericardial resection disclosing the spread of the tumor to both ventricle walls.

right knee in the form of a slow-growing soft tissue mass.

Clear cell sarcoma may follow various clinical courses. The diameter of these tumors is considered to be a very significant factor related to survival.[3] Patients with tumors having a diameter larger than 5 cm, as in our case, have a low rate of survival. Recurrence did not occur in our patient, but distant metastasis appeared 14 months after her diagnosis. The incidence of local recurrence and distant metastasis have been reported as 14-26% and 44-83%, respectively.[1,3] Eckardt et al.[4] reported that local lymph node metastasis and local recurrence were predictors of a poor prognosis. We did not detect local recurrence and lymph node metastasis in our patient; however, cardiac metastasis was seen, and that affected the prognosis unfavorably.

A histopathological examination of the tumor showed necrotic areas. Lucas et al.[3] reported that the presence of necrosis in histopathological sections is an independent factor for the poor prognosis in clear cell sarcoma. Our findings correlated with that report.

A wide surgical excision of the tumor, local adjuvant radiotherapy, and chemotherapy are available treatment alternatives for clear cell sarcoma. The adjuvant radiotherapy is useful for local control of this type of cancer in cases where the surgical borders are suspicious.[4] The effectiveness of adjuvant chemotherapy is not clear. Recurrences following doxorubicin-based chemotherapy have been reported for soft tissue sarcoma that is locally confined.[5]

Chemotherapy is reported to be more useful in cases with a disseminated tumor.[2] Our patient lived for 14 months without any symptoms or signs of recurrence after chemotherapy. Steger et al.[6] reported that complete resolution can be obtained after administration of interferon-alpha 2b (IFN-α 2b), but there are no other reports that confirm this. In addition, studies are underway to recognize tumor-specific antigens as targets in tumor cells for successful immunotherapy.[7]

The option of a radical tumor excision offered to the patient could not be carried out because she adamantly refused any further surgical treatment. This also ruled out any possibility of planning for cardiac transplantation, which is also relatively contraindicated in patients with a recent malignancy.[8]

Our case showed that clear cell sarcoma behavior in the heart is aggressive. In our patient, a wide excision of the tumor was carried out as the primary surgical treatment. The patient's therapy was complemented by local adjuvant radiotherapy followed by chemotherapy at a later date. Cardiac metastasis was detected fourteen months after diagnosis. Repeat pericardial effusions in our patient led to palliative surgical intervention to open a pericardial window. Ultimately, this disease has a poor prognosis when there is metastasis to cardiac tissues, and this has also been previously reported in other metastatic clear cell sarcomas.

Herein, cardiac metastasis of clear cell sarcoma with pathological confirmation was reported for the first time, and it proved to be catastrophic in our patient. Although opening a pericardial window and performing pericardial resection provided temporary relief, these procedures had no effect on the clinical course. The patient's prognosis remained poor due to the rapid growth of the tumor and she failed to survive an emergent median sternotomy undertaken to due to cardiac arrest.

Declaration of conflicting interests

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REFERENCES

1. Enzinger FM. Clear-cell sarcoma of tendons and aponeuroses. an analysis of 21 cases. Cancer 1965;18:1163-74.
2. Kuiper DR, Hoekstra HJ, Veth RP, Wobbes Th. The management of clear cell sarcoma. Eur J Surg Oncol 2003;29:568-70.
3. Lucas DR, Nascimento AG, Sim FH. Clear cell sarcoma of soft tissues. Mayo Clinic experience with 35 cases. Am J Surg Pathol 1992;16:1197-204.

4. Eckardt JJ, Pritchard DJ, Soule EH. Clear cell sarcoma. A clinicopathologic study of 27 cases. Cancer 1983;52:1482-8.
5. Adjuvant chemotherapy for localised resectable soft-tissue sarcoma of adults: meta-analysis of individual data. Sarcoma Meta-analysis Collaboration. Lancet 1997;350:1647-54.
6. Steger GG, Wrba F, Mader R, Schlappack O, Dittrich C, Rainer H. Complete remission of metastasised clear cell sarcoma of tendons and aponeuroses. Eur J Cancer 1991;27:254-6.
7. Liao SK, Perng YP, Lee LA, Chang KS, Lai GM, Wong E, et al. Newly established MST-1 tumour cell line and tumour-infiltrating lymphocyte culture from a patient with soft tissue melanoma (clear cell sarcoma) and their potential applications to patient immunotherapy. Eur J Cancer 1996;32A:346-56.
8. Nwakanma LU, Shah AS, Conte JV, Baumgartner WA. Heart transplantation. In: Cohn LH, editor. Cardiac surgery in the adult. New York: McGraw-Hill; 2008. p. 1539-78.