Rijetki slučaj metastatskog pleuropulmonalnog sinovijalnog sarkoma s opstrukcijom mitralnog zaliska: prikaz slučaja

Rare case of metastatic pleuropulmonary synovial sarcoma causing mitral valve obstruction: a case report

Mijo Meter*, **Zora Sušilović Grabovac**, **Ivica Vuković**, **Ivan Gudelj**, **Nikola Crnčević**, **Frane Runjić**

Klinički bolnički centar Split, Split, Hrvatska

University Hospital Centre Split, Split, Croatia

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***ADDRESS FOR CORRESPONDENCE:** Mijo Meter, Klinički bolnički centar Split, Spinčićeva 1, HR-21000 Split, Croatia. / Phone: +385-98-9752-946 / Email: mijometer05@gmail.com

ORCID: Mijo Meter, https://orcid.org/0000-0003-4674-426X • Zora Sušilović Grabovac, https://orcid.org/0000-0001-9999-7557
Ivica Vuković, https://orcid.org/0000-0003-2170-1327 • Ivan Gudelj, https://orcid.org/0000-0001-9314-7164
Nikola Crnčević, https://orcid.org/0000-0002-1399-3406

Uvod: Pleuropulmonalni sinovijalni sarkomi rijetki su zloćudni tumori mekog tkiva.[1-3] No, metastaza pleuropulmonalnog sinovijalnog sarkoma s valvularnim lezijama, što je bio slučaj s našom bolesnicom, još je rjeđa. Predstavljamo slučaj 42-godišnje bolesnice s metastatskim pleuropulmonarnim sinovijalnim sarkomom koji se proširio u šupljinu lijeve klijetke i prouzročio opstrukciju mitralnog zaliska.

Prikaz slučaja: 42-godišnja pacijentica s anamnezom reseciranog ingvinalnog sinovijalnog sarkoma koja je bila podvrgnuta radikalnom zračenju i ostala u remisiji iduće tri godine javila se sa sljedećim simptomima: kašljem, dispnejom i hemoptizama. Snimka kompjutorizirane tomografije (CT) ukazala je na tumorsku masu u donjem lijevom plućnom režnju koja je pritiskala medijastijalnu pleuru s infiltracijom lijeve plućne vene. CT je također otkrio masu od 6 cm u lijevoj pretklijetki koja se širila u mitralni zalistak. Urađena je hitna ehokardiografija koja je otkrila masu od 60x27 mm u lijevoj pretklijetki sa širenjem u mitralni zalistak koja je prouzročila valvularnu opstrukciju. Srednji gradijent tlaka preko mitralne valvule iznosio je 6 mmHg. Ehokardiografija je prikazala urednu ventrikularnu sistoličku funkciju, ali i proširenu desnu klijetku s teškom plućnom hipertenzijom. MRI je potvrdio prethodne rezultate, no pokazao je značajni prodor tumorske mase u desnu pretklijetku kroz atrijalni septalni defekt. Bronhijalna biopsija tumorske mase otkrila je nediferencirane monomorfne svijetle vretenaste stanice s pozitivnom reakcijom na CD99 čime je potvrđena dijagnoza metastatskog monofaznog sinovijalnog sarkoma. Zbog velikog proširenja tumora na gotovo sve srčane šupljine, daljna kardiokirurška obrada nije bila indicirana. Pacijentica se sada upućuje na daljnju kemoterapiju ifosfamidom/doksorubicinom.

Zaključak: Agresivna priroda sinovijalnog sarkoma otežava njegovo rano otkrivanje. Zbog malog broja zabilježenih slučajeva, nema konsenzusa oko najučinkovitije terapije. Stoga se čini korisnim istražiti najučinkovitije kirurške postupke i dodatne protokole kemoterapije radi povećanja stope preživljavanja pacijenata.

Introduction: Pleuropulmonary synovial sarcomas are rare soft tissue malignancies.[1-3] Additionally, pleuropulmonary synovial sarcoma metastasis with valvular involvement, which was the case with our patient, is even more rare. We present an unusual case of a 42-year-old female patient with metastatic pleuropulmonary synovial sarcoma prolapsing to the left ventricular cavity and causing mitral valve obstruction.

Case report: 42-year-old female patient with a history of previously resected inguinal synovial sarcoma who underwent radical irradiation and remained stable for 3 years appeared with the following symptoms: cough, dyspnea and hemoptysis. Computed tomography (CT) scan showed a suspected tumor mass within the left inferior pulmonary lobe affecting the mediastinal pleura with infiltration of the left pulmonary vein. CT scan also revealed a 6 cm mass in the left atrium (LA) that prolapsed into the mitral valve. Immediate echocardiography was performed, which showed a left atrial mass measuring 60x27mm prolapsing into the mitral valve and causing valvular obstruction. Mean pressure gradient across the mitral valve was 6 mmHg. Echocardiography revealed normal ventricular systolic function but a dilatated right ventricle with severe pulmonary hypertension. MRI confirmed previous echo findings but showed a more prominent protrusion of the tumor mass to the right atrium through the atrial septal defect. Bronchial biopsy of the tumor mass revealed undifferentiated monomorphic blunt spindle cells with CD 99 positive expression which confirmed the diagnosis of metastatic monophasic synovial sarcoma. Due to a large tumor extension to almost all cardiac chambers, cardiothoracic surgery was not indicated. Patient is considered for further chemotherapy with ifosfamide/doxorubicin.

Conclusion: The aggressive nature of synovial sarcoma makes its early detection difficult. Because of the small number of reported occurrences, there is no consensus regarding optimal therapy. Therefore, it seems worth investigating both optimal surgical procedures and additional chemo-radiotherapy protocols in order to improve the patient's survival.

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