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CASE REPORT

Acute left and right ventricular heart failure with transient myocardial edema in a 26-Year-old female patient with peripartum cardiomyopathy and clivus chordoma

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Abstract

Peripartum cardiomyopathy (PPCM) is a life-threatening disease characterized by the clinical symptoms of acute heart failure with reduced ejection fraction in women at the end of their pregnancy or in the first months after delivery. A 26-year-old woman was delivered by cesarean section at 29 weeks due to increasing intracranial pressure from a newly diagnosed clivus chordoma. She was transferred to ICU after septic deterioration, presenting with hemodynamic instability, tachycardia, high catecholamine support, oxygen needs, and signs of cardiac decompensation. We found a significant impairment of the left and as well as the right ventricular function (LVEF 30–35%, FAC 30%) combined with pericardial effusion and a septal myocardial edema (cardiac MRI). There were no signs of myocardial inflammation and no hint for a Takotsubo cardiomyopathy neither but in total pointing to a PPCM. Left and right cardiac function improved stepwise after breastfeeding was stopped and bromocriptine was started, supporting our theory that the patient suffered from a peripartum cardiomyopathy involving both ventricles. Although right ventricular function was also reduced, which is a marker of a severe process, cardiac function improved quickly. This case shows that even in complex intensive care cases in postpartum patients, a cardiac genesis of the hemodynamic deterioration should always be considered and clarified to be able to provide these patients with adequate therapy and regulate the cardiac course of PPCM.

Keywords Peripartum cardiomyopathy · Clivus chordoma · Reduced left and right ventricular function · Heart failure

Abbreviations

BSA	Body surface area
EDV	Enddiastolic volume
ESV	Endsystolic volume
FAC	Fractional area change
GDMT	Guideline-directed medical therapy
HFrEF	Heart failure with reduced ejection fraction
hs-TNI	High-sensitivity troponin I
IVSd	Interventricular septum thickness at end-diastole
LVEF	Left ventricular ejection fraction
PPCM	Peripartum cardiomyopathy
PEG	Percutaneous endoscopic gastrostomy
sFLt 1	Soluble fms-like tyrosine kinase 1

Background

Peripartum cardiomyopathy (PPCM) is characterized by new-onset cardiomyopathy with an impaired left ventricular ejection fraction less then at least 50% in women towards the end of their pregnancy or within the months following delivery, abortion or miscarriage, without any other causes for heart diseases such as preexisting conditions [1, 2]. Clinical manifestations range from mild signs of cardiac decompensation up to potentially life-threatening complications with acute heart failure [2–5]. Due to often non-specific

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clinical presentation as well as the coexistence of fatigue, leg edema or shortness of breath as part of the hemodynamic adaptation mechanisms of a normal pregnancy, the awareness of this potential diagnosis in pregnant and peri/post-partal females is essential for an early detection and therapy [2, 3]. PPCM is associated with high morbidity and mortality, but timely diagnosis can significantly improve the chances of full recovery [5].

Important differential diagnoses such as hypertensive pregnancy diseases, pulmonary artery embolism and cardiac ischemia must be ruled out for the diagnosis of PPCM [2, 3, 5]. Cardiac biomarkers, in particular natriuretic peptide, and basic cardiac diagnostics using ECG and echocardiography are useful for diagnosis [2, 5]. The pathogenesis of PPCM is multifactorial and not completely understood [2, 6]. Heterogenous hypotheses involve angiogenic, hormonal and metabolic factors, which lead to PPCM in combination with predisposition [2, 6–8]. There is a hint that a PPCM occurs more often in females that have survived cancer—most likely due to the pre-existing cardiac damage [9].

Treatment of peripartum cardiomyopathy is largely limited to the same neurohormonal antagonists used for other forms of heart failure and cardiomyopathy. There are still no proven disease-specific therapies. Research over the past decade suggests that peripartum cardiomyopathy is caused by vascular dysfunction triggered by maternal hormones in late gestation. Recently, there is also evidence that many cases of peripartum cardiomyopathy are genetic. Based on the case described, we classify the disease in the known epidemiology and give an overview of the treatment of peripartum cardiomyopathy [10].

Various factors and image morphological changes such as reduced left ventricular as well as right ventricular function are prognostically relevant for the course of the disease [11]. Thanks to the interdisciplinary care of our patient and the availability of echocardiography and cardiac magnetic resonance imaging (MRI), we were able to reconstruct the course of the disease appropriately.

Case presentation

A 26-year-old woman was delivered by cesarean section at 29 weeks due to increasing intracranial pressure from a newly diagnosed, slowly growing, locally aggressive clivus chordoma. The tumor's progression and neurological deficits led to the pregnancy's termination to enable prompt surgical treatment of the brain tumor. In the postoperative course, the young patient developed fever, circulatory instability and signs of septic shock with several complications and further computed tomography (CT) imaging was performed. This revealed pneumonia and as leading cause of the septic shock a 4-quadrant peritonitis due to an insufficient percutaneous endoscopic gastrostomy (PEG) tube. Empiric antibiotic therapy with piperacillin/tazobactam was initiated, followed by escalation to Meropenem due to lack of clinical response. Intraoperative cultures obtained—during exploratory laparotomy performed for the treatment of the four-quadrant peritonitis—revealed the presence of *Klebsiella oxytoca*. The CT showed as incidental findings a cardiomegaly with pericardial effusion. As part of the septic course of the disease, the patient was initially transferred to the intensive care unit. During the intensive care stay, the patient was then presented to cardiology for the first time. Echocardiographic findings (please see supplementary video 1 and 2) revealed a pericardial effusion, septal myocardial edema and signs of cardiac decompensation with significant impairment of left and right ventricular function with a LVEF of 30–35% and a reduced right ventricular function (FAC 30%, TAPSE 25 mm). In particular, the septal myocardial edema was impressive in the echocardiographic images (please see supplementary video 1 and 2). There was a significant left ventricular and interestingly also a right ventricular edema, raising the possible differential diagnosis of septic cardiomyopathy or a pre-existing cardiomyopathy. The interventricular septum was up to 15 mm thick (please see Fig. 1). However, there were no indications of congenital diseases at the time of presentation.

The clinical examination revealed an invasively ventilated, highly catecholamine-dependent and clinically decompensated patient with pleural effusions, pericardial effusion and leg edema. Blood samples showed elevated inflammation parameters (CRP 310 mg/dl, procalcitonin 11 ng/ml, white blood cell count 13.7/nl) consistent with the septic course of the disease as well as significantly elevated cardiac biomarkers with an NT-proBNP of 26,369 pg/ml and a low elevated high sensitivity-TNI of 53 pg/ml (see Fig. 2).

During intensive care stay and the patient's clinical deterioration, the pumping of breast milk was stopped, and breastfeeding was ended. The treatment of heart failure was initiated despite persistently low blood pressure, initially with a low-dose beta blocker (bisoprolol 2.5 mg/day), ACE inhibitor (ramipril 2.5 mg/day) and an SGLT-2 receptor antagonist (empagliflozin 10 mg/day).

A few days after initiation of adequate guideline-directed medical therapy (GDMT) for heart failure medication and supplemented by intravenous diuretics, a reduction in these edematous components was observed. After stabilization of the patient, removal of the cerebral shunt and establishment of MRI suitability, cardiac MRI imaging was performed. Findings in cardiac MRI showed an enlarged heart (EDV 173 ml, ESV 112 ml, EDV/BSA 112ml/m^2, ESV/BSA

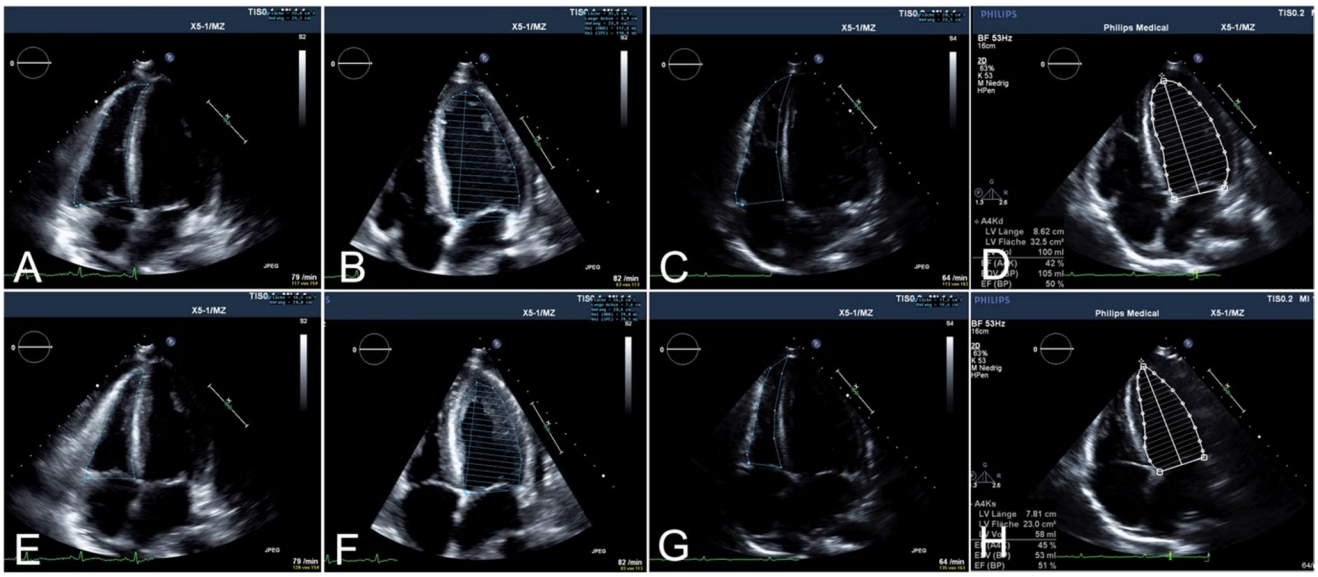

Fig. 1 Echocardiographic findings before and after initiation of GDMT with focus on right and left ventricular functional findings. **A**, **E** Right ventricular function at manifestation and before initiation of GDMT with FAC 30%, **B**, **F** left ventricular function at manifestation and before initiation of GDMT with biplane EF 33%, **C**, **G** Right ventricular function after initiation of GDMT with FAC 60%, **D**, **H** Left ventricular function after initiation of GDMT with biplane EF 51%

72ml/m^2) with septal hypertrophy accentuated basal to mid-ventricular and with normalization towards the apex.

Quantitative tissue characterization by T1—(1455ms) and T2—(48ms) mapping revealed increased relaxation times of the septal myocardium without any focal late enhancement (see Fig. 3). Myocarditis, Tako-Tsubo cardiomyopathy or septic cardiomyopathy was ruled out and the initial suspicion of peripartum cardiomyopathy was strengthened. The left ventricular pump function was also reported as 36% on MRI. The septal myocardial edema showed in repeated echocardiography a clear regression in the short term with a reduction of the interventricular septum thickness an end-diastole (IVSd) from 15 mm to 11 mm (please see supplementary video 3 and 4). After excluding further cardiac causes, bromocriptine therapy as a targeted therapy for PPCM was initiated. Further close monitoring under adequate heart failure medication showed a stabilization of left ventricular systolic ejection function (LVEF 50%), right ventricular function (FAC 60%, TAPSE 25 mm) and a significant decrease in cardiac biomarkers (NT-proBNP 2815 pg/ml, hs-TNI 10pg/ml) as well (please see supplementary video 3 and 4).

In the following weeks, the underlying disease progressed with an infiltrative growth of the clivus chordoma, so that the patient's prognosis had to be re-evaluated as unfavorable. The treatment was changed to best supportive care and palliative medical co-care as well as palliative radiation of the tumor to give the young family some time.

Several limitations of the course need to be critically discussed: The initial cardiac decompensation and diagnosis of reduced cardiac ejection function occurred at the same time as the first septic deterioration with 4-quadrant peritonitis. The differential diagnosis of septic cardiomyopathy could never be completely ruled out. When the patient was transferred to the intensive care unit, her weaning from breastfeeding was also noted, leading to the suspicion that peripartum cardiomyopathy was likely. The primary treatment of the sepsis was uncomplicated, but the cardiac abnormalities, particularly the moderately-to-severe reduced ejection function, pericardial effusion and myocardial edema persisted well beyond the critical episode and the intensive care course. With the use of adequate heart failure therapy and intravenous recompensation, a clinical and morphologic improvement appeared. Following a recurrent septic deterioration necessitating multiple abdominal surgeries for the management of the primary focus of peritonitis (see days 41–46 in Fig. 2), the previously improved cardiac function remained stable and continued to recover despite the renewed septic course. Echocardiographic control after initiation of bromocriptine therapy was finally not performed as the patient was transferred to palliative care as the tumor rapidly progressed.

Discussion and conclusions

Postpartum cardiomyopathy is a type of (left and right ventricular) heart failure that manifests itself at the end of pregnancy or in the puerperium [1–4, 7, 12]. Especially the involvement of the right ventricle in particular can indicate a

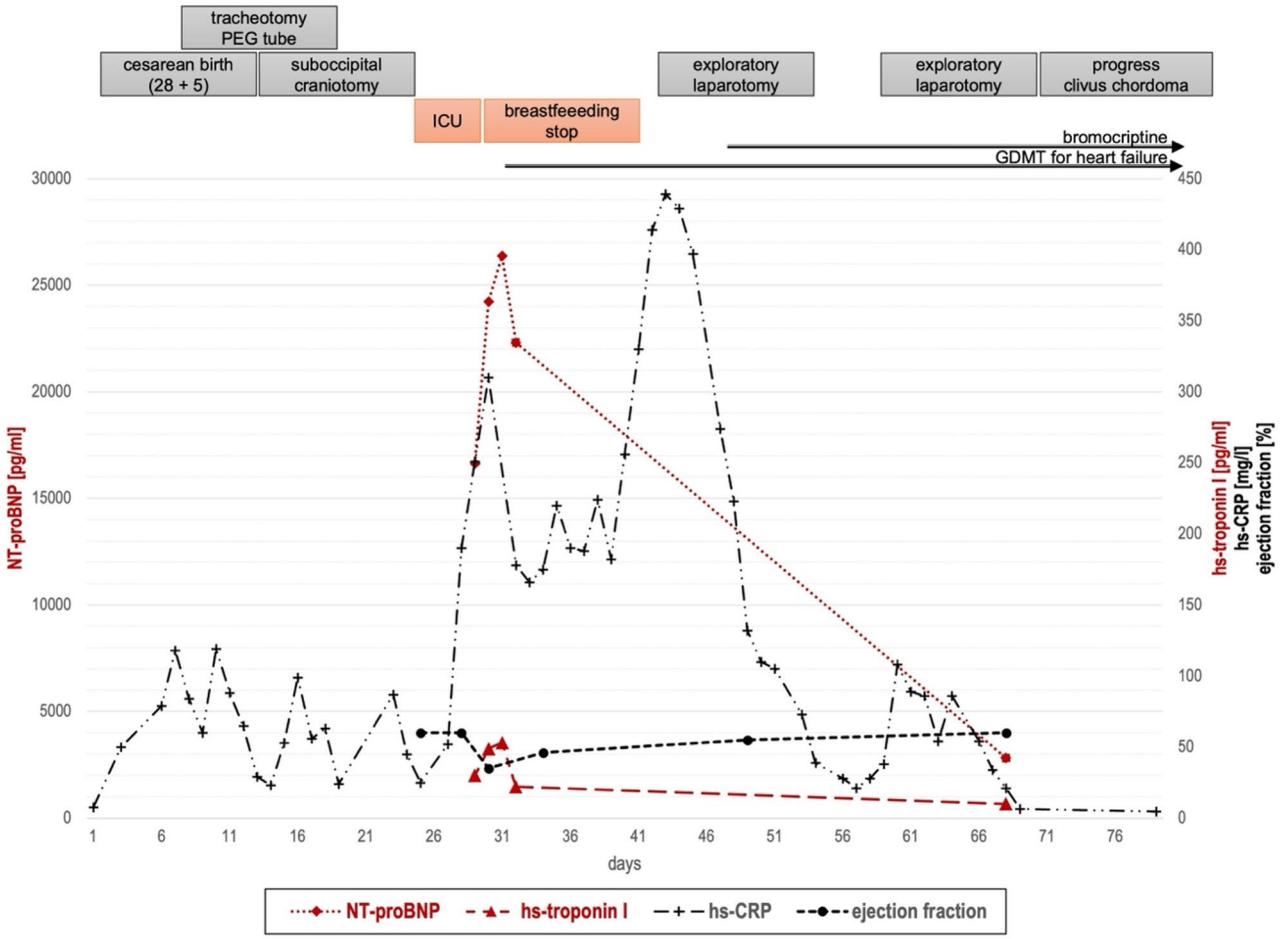

Fig. 2 Clinical course with graphical representation of cardiac biomarkers NT-proBNP and hs-troponin I (red) and an inflammatory biomarker (hs-CRP, black) to demonstrate the septic course and the occurrence of PPCM. The days since admission with initial cesarean birth, tracheotomy and PEG placement for subsequent tumor resection by suboccipital craniotomy are shown. Cardiomyopathy was diagnosed on the transfer to the intensive care unit; after breastfeeding stop and the start of guideline-directed medical therapy (GDMT), the cardiac biomarkers showed a decreasing trend

severe course of PPCM [13]. The pathogenetic mechanisms, risk factors and potential clinical course of the disease are repeatedly focus of scientific analysis and investigation and still incompletely understood [1–4, 7, 12].

Important maternal risk factors identified so far include smoking, diabetes, preexisting pre-eclampsia, African ethnicity, advanced maternal age or teenage pregnancy, lack of formal education, unemployment and underweight status [1, 5, 12]. Other less studied risk factors relate to the pathogenetic mechanisms and changes during pregnancy, which in combination seem to contribute to developing PPCM [1, 5]. These include activation of inflammatory pathways, the impact of prolactin metabolites, which could induce vasoconstriction, apoptosis and therefore inflammation as well as capillary damage and underlying genetic predisposition like biologically active 16kDA prolactin, soluble fms-like tyrosine kinase 1 (sFlt 1) [5, 8]. Important in clinical daily life is to have this rare cause of heart failure in mind as potential differential diagnosis in females that are pregnant or have just given birth to a child– to pave the way for the correct therapy.

Even if a septic component could not be definitively ruled out in our patient as cause of heart failure the course of the disease gives strong hint for an underlying of PPCM: the cardiomyopathy manifested itself immediately peri-/postpartum, improved after breastfeeding was stopped and the patient benefited clinically and hemodynamically from bromocriptine therapy. On the other side the diagnosis of HFrEF was made upon admission to the intensive care unit in the context of septic shock. Septic cardiomyopathy is one common complication and could initially have been considered a plausible explanation for the patient's clinical course. However, despite adequate anti-infective therapy, of breastfeeding and milk expression, a rapid clinical improvement was observed within a few days. Further improvement occurred after initiation of heart failure therapy. In summary

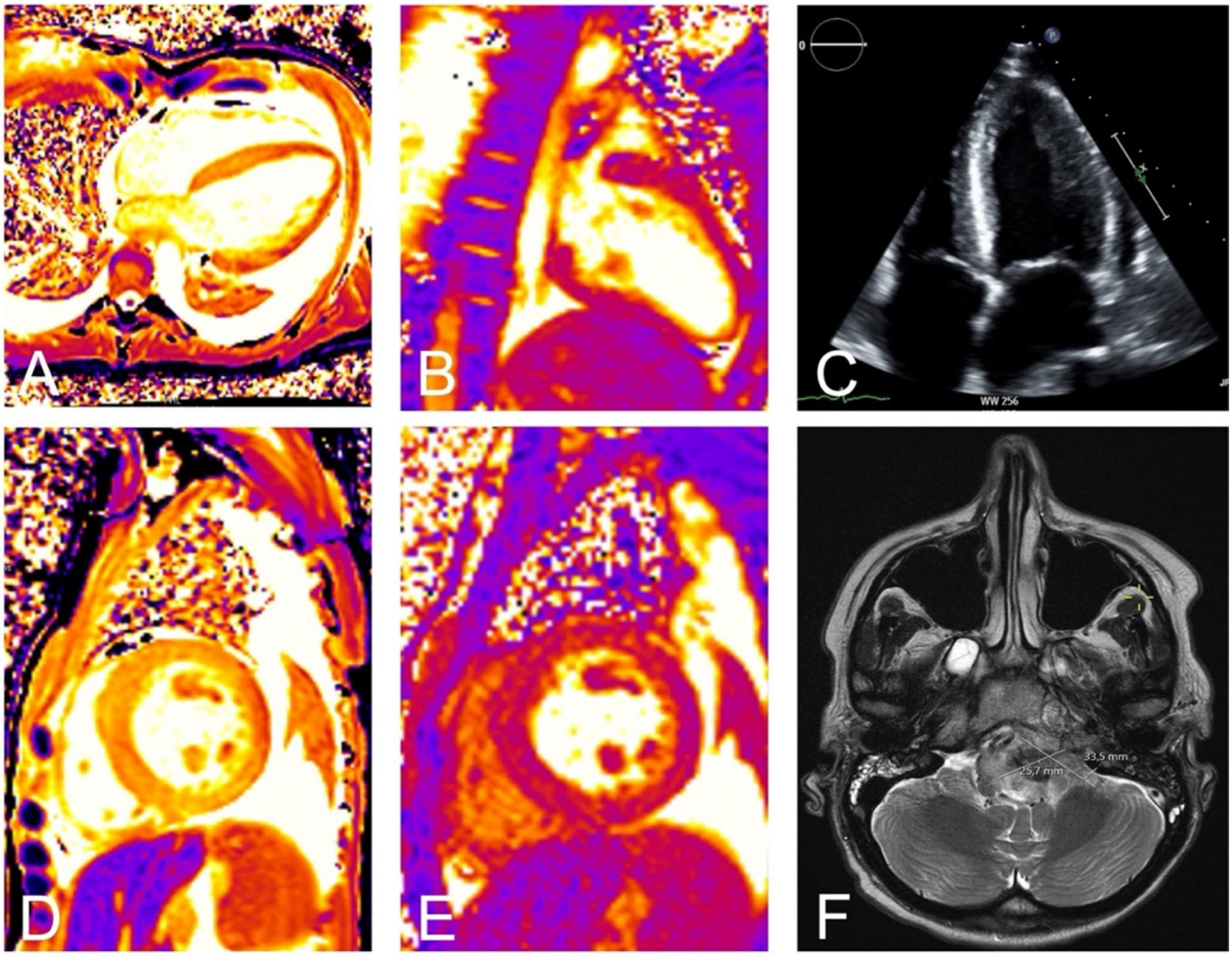

Fig. 3 Cardiac MR, echocardiography and cerebral CT of the patient. Enlarged left ventricle (EDV 173 ml, ESV 112 ml, EDV/BSA 112ml/m^2, ESV/BSA 72ml/m^2) with septal hypertrophy accentuated basal to midventricular and with normalization towards the apex with increased relaxation times of the septal myocardium in the **A** and **D** T1—(1455ms) and **B** and **E** T2—weighted (48ms) sequences without any focal late enhancement; **C** Enddiastolic 4-chamber view with pericardial effusion, moderately to severe reduced left and right ventricular function and hypertrophic septum; **F** Clivus chordoma with intracranial infiltration shown by MRI.

of the potential clinical differential diagnoses and based on close echocardiographic and laboratory monitoring, we consider peripartum cardiomyopathy (PPCM) to be more likely than septic cardiomyopathy. MRI morphology was able to rule out other causes of cardiomyopathy such as an inflammatory cardiomyopathy and fibrosis formation.

The association of PPCM with history of cardiotoxic therapy for a malignant disease is known and has been described [9]—but this cannot be the case in our patient presented here: The malignant diagnosis of clivus chordoma was made during pregnancy, primary therapy was tumor resection and no chemotherapy was given. So, there is no cancer therapy associated component of the cardiomyopathy in the presented patient.

Several clinical and laboratory findings such as the severely reduced left ventricular function combined with left ventricular dilatation and especially the reduced right ventricular function and are associated with prolonged disease progression and worse outcome in PPCM [5, 11, 13]. Interestingly, the right ventricular function of the patient recovered quickly and completely after breastfeeding was stopped and bromocriptine was started, in parallel with the improved left ventricular function.

In general, diagnosis and optimal treatment of women with postpartum cardiomyopathy requires a multidisciplinary team with expertise in the management of high-risk pregnancies and heart failure [5, 14]. Treatment options for postpartum cardiomyopathy include drug therapy with heart failure medication, bromocriptine as a dopamine antagonist and anticoagulants. Bromocriptine is a well-established approach in the therapy for postpartum cardiomyopathy

because it suppresses the release of prolactin, thereby providing potential benefits [5, 14].

Our reported case demonstrates PPCM as a severe complication of pregnancy and post-pregnancy with a life-threatening course due to acute heart failure - associated with the malignant disease of a clivus chordoma. To our knowledge, this is the first report of a case of PPCM in a patient with a malignant tumor and, on top, a concomitant septic disease course. As the potential diagnosis of the PPCM was early made and the correct therapeutical consequences were put into practice, the cardiological situation improved markedly despite the right ventricular involvement marking a severe course.

The clinical course of our patient presented here was nevertheless intriguing due to the life-limiting comorbidities. Based on the hypothesis that pregnancy may have led to downregulation of immunologic processes and therefore might have favored the development of the PPCM and potentially also of the clivus chordoma, it remains unclear what factors may have further favored one or the other.

However, our case report shows once more the heterogeneous genesis of PPCM and the importance to have this differential diagnosis in mind, but also the importance of further (basic) research and studies on PPCM, as the entity is still not completely understood. A promising registry study, the EURObservational Research Programme international, will be able to provide more information about PPCM with basic data in the next few years and address some of the remaining questions [6].

In general, the awareness that the PPCM could be the underlying cause for heart failure in pregnant/post-pregnant women and the correct therapeutic consequences (stop breastfeeding, bromocriptine and adequate heart failure medication) is important and helps to improve the cardiological outcome. Nevertheless, in our case reported here, the malignant co-existing disease formed the life-limiting aspect.

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Author contributions Material preparation, data collection and first draft of the case report were performed by SK. All authors read and approved the final manuscript.

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Data availability All data generated or analyzed during this study are included in this published article.

Declarations

Competing interests SK received speaker honoraria from AstraZeneca not associated with this work. SHK declares having received consultancy honoraria from Almirall and lecture honoraria from Janssen-Cilag GmbH not associated with this work. All other authors declare that they have no conflicts of interest in the context of this work.

Ethical approval and consent to participate Informed consent was obtained from the patient.

Consent to publication Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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