Management of Leukemia and Partial Atrioventricular Septal Defect during Pregnancy

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Abstract

Keywords
► minimally invasive surgery
► congenital heart disease
► CHD
► hematology
► pregnancy

Background Pregnancy-associated acute myeloid leukemia (PA-AML) is rare. Cardiac surgery in the context of AML poses challenges that are seldom encountered.

Case Description The subject is a 31-year-old woman at 38 weeks’ gestational age diagnosed with AML and partial atrioventricular septal defect. After multidisciplinary consulting, an urgent cesarean section was performed, then chemotherapy was initiated, followed by minimally invasive cardiac surgery with an uneventful recovery.

Conclusion Efficient multidisciplinary approach is essential in the management of PA-AML and cardiac disease. Minimally invasive cardiac surgery may be safe and useful in patients with AML.

Introduction

Acute myeloid leukemia (AML), generally occurring in older adults, has been reported in 1 out of 75,000 to 100,000 pregnancies.1 Current data on clinical features and treatment of pregnancy-associated AML (PA-AML) are scarce.1 Complicating 1% of pregnancies, cardiovascular disease is the leading cause of indirect maternal mortality in developed countries.2 Although adult congenital heart disease (CHD) is the most common type of heart disease encountered during pregnancy, partial atrioventricular septal defect (PASVD) is rare.2 Herein, we describe a case with PA-AML and PASVD who was successfully treated with multidisciplinary treatment including an urgent cesarean section (CS), chemotherapy, and a minimally invasive cardiac surgery.

Case Description

A 31-year-old Chinese woman at 38 weeks’ gestational age was referred to our hospital for progressive dyspnea on exertion and rapid decrease of peripheral blood counts over a 2-month period. The patient had a previous abortion 7 years ago. On examination, she had an elevated respiratory rate of 30 breaths/min and a pulse oximetry of 91%, and was normotensive (120/88 mm Hg) and tachycardiac (HR 120 bpm) with precordial systolic murmur. Her lower extremities were edematous. Her obstetric examination findings and fetal sonography results were normal and appropriately correlated with the gestational age. Peripheral blood counts showed white blood cell 6.22 × 10^9/L, hemoglobin 88 g/L, and platelets 46 × 10^9/L. Echocardiography demonstrated a large ostium primum atrial septal defect (2.1 cm) with significant shunt, cleft left atrioventricular valve with severe insufficiency, patent foramen ovale (PFO) (< Fig. 1A, B), and moderate tricuspid regurgitation with an ejection fraction of 66%. The pulmonary artery pressure was estimated to be 43 mm Hg.

After multidisciplinary consulting, an urgent CS was performed under general anesthesia without complications and a morphologic normal female weighing 3,100 g was born. The mother’s bone marrow aspirate revealed a diagnosis of AML. Fluorescence in situ hybridization karyotyping was positive for t(8:21) (q22;q22). After the CS, the patient’s vital signs and...
oxygenation gradually returned to normal values. Then induc-
tion chemotherapy was initiated, a complete remission
obtained, followed by two cycles of consolidation chemother-
apy. Subsequently video-assisted cardiac surgery was per-
formed to correct her cardiac defects. We used a 5-cm right
submammary groove approach with a 5-mm endoscope to
enhance intracardiac visualization. Peripheral cannulation
was used for cardiopulmonary bypass. A vertical right atriot-
omy incision was used to expose and incise the septum
primum, which allowed repair of the left atrioventricular
valve. The cleft was closed with a continuous 5/0 Prolene
suture, and the ostium primum defect was repaired with the
use of an autologous pericardial patch (►Fig. 2). PFO was
closed with a horizontal mattress pledgeted 4/0 Prolene
suture. A Sorin 28-mm ring was implanted for tricuspid
annuloplasty. No blood product was given perioperatively.
Postoperative echocardiography demonstrated no residual
shunt and trace mitral insufficiency (►Fig. 1C, D). The patient
was extubated in the sixth hour after surgery and discharged
home without complication after 9 days in hospital. The
patient and the infant are alive and in good health after a
follow-up of 12 months.

**Discussion**

Signs and symptoms experienced during a normal pregnancy
are similar to those of cardiac disease, leading to a confusing
clinical picture. The clinical feature of AML presenting during
pregnancy is also similar to that of nonpregnant women. Thus,
the diagnosis can be easily delayed. In our case, the
patient lived in a remote area. Her CHD was unnoticed until her
visit to our hospital. The diagnosis of PA-AML and PAVSD was
confirmed very late (at 38 weeks’ gestational age), partly
because her symptoms were nonspecific.

Preexisting cardiac conditions can interact with the physio-
logical changes occurring during pregnancy and increase the
risk of maternal and fetal adverse outcomes. Our patient had
hepatomegaly and edematous lower extremities, and the echo
showed a dilated right ventricle and atrium with elevated
pulmonary systolic pressure, which confirms the diagnosis of
right heart failure. Acute leukemia may present with hyper-
leukocytosis, thromboses, or disseminated intravascular coag-
ulation, in the context of a gestational associated thrombogenic
milieu. Hemorrhage secondary to acute leukemia is also
common. Thus, the treatment of PA-AML and PAVSD is
extremely challenging. Our patient fell into extremis, develop-
ing right heart failure. Taking into account the immediate threat
to the lives of the woman and fetus, emergent CS was promptly
accomplished. For PA-AML, chemotherapy remains the main-
stay of treatment. Based on the available data, PA-AML treated
with appropriate regimens might obtain outcomes similar to
those of nonpregnant woman, and the prognosis is worse only
when appropriate treatment is significantly delayed. In our

![](image.png)

**Fig. 1** Preoperative TTE showed (A) an ostium primum ASD and PFO, and (B) severe left-sided atrioventricular valve insufficiency. Postoperative TEE confirmed an intact septum without residual shunt (C) and trace mitral insufficiency (D) after surgery. ASD, atrial septal defect; LA, left atrium; LV, left ventricle; PFO, patent foramen ovale; RA, right atrium; TTE, transesophageal echocardiography.
the potentially fatal uterine bleeding associated with cardio-
pulmonary bypass and the delay to initiate chemotherapy
which would negatively affect the mother’s prognosis.

To our knowledge, this is the first case of PAVSD in PA-AML
with subsequent minimally invasive cardiac surgery. AML is
characterized by uncontrolled proliferation of immature,
abnormal blast cells, and impaired production of normal blood
cells.\(^1\)\(^4\) Surgical trauma and cardiopulmonary bypass, because of
their immune-depressing effects, have the potential risk of
increasing the hematological problems, leading to fatal or
morbidity complications.\(^5\) The current mortality risk calculators
used for cardiac surgery do not account for hematological
malignancy, but several studies have concluded that patients
with hematological malignancy carry an increased risk of
complications.\(^5\) To reduce the risk of postoperative infection
and thrombohemorrhagic complications, we performed the
cardiac surgery after obtaining complete remission and
we waited until two cycles of consolidation treatment were
done to confirm that there was no relapse.

Because of its good exposure, median sternotomy has been
the conventional approach for cardiac surgery. However, it
causes long-term adverse effects on patients with large trau-
ma, bleeding, long-term hospitalization, and obvious scars
after recovery. Recently, minimally invasive surgery has gained
popularity as it is associated with decreased blood loss, less
postoperative pain, shorter hospitalization time, faster recov-
ery, and better cosmesis.\(^6\) Minimally invasive cardiac surgery
is also related to reduced inflammatory reaction and coagu-
lopny.\(^7\) It is thus believed that high-risk patients benefit most
from minimally invasive surgery. Our AML patient clearly
derived healing benefits from minimally invasive surgery,
avoiding issues with sternal healing and postoperative infec-
tions. Totally robotic repair of PAVSD has also been reported,\(^8\)
but the high expenses associated with it cannot be overlooked.
The long operation time was thought to be a shortcoming of
minimally invasive cardiac surgery. However, the operative
duration can be significantly shortened with surgical experi-
ence gained.\(^8\)

**Conclusion**

In summary, this is a rare case of the very unusual coinci-
dence of PA-AML and PAVD at full-term pregnancy. Efficient
multidisciplinary approach is essential in management. Minimally invasive surgery may be safe and useful in patients
with AML.

**Conflict of Interest**

None declared.

**References**

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