Cyanotic Tetralogy of Fallot With Its Infective Endocarditis Complication on the Tricuspid and Pulmonary Valves of a 55-Year-Old Man

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A 55-year-old man had undiagnosed tetralogy of Fallot with the complications of decompensated heart failure and infective endocarditis, as well as pulmonic involvement secondary to the endocarditis. The patient had a massive hemoptysis and died. This case is a rare insight into the late outcome of this congenital heart disease. (Circ J 2004; 68: 178–180)

Key Words: Infective endocarditis; Middle age; Tetralogy of Fallot

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease. Survival into adulthood is possible, but uncommon without surgical intervention and less than 5% of patients survive to age 40. Infective endocarditis (IE) is a serious and fatal complication in adults with congenital heart disease. We report a man who survived to the age of 55 years with undiagnosed cyanotic TOF, complicated by decompensated heart failure and IE. The case highlights some difficult problems encountered in the middle-aged patient with uncorrected TOF, a situation in which the medical literature provides little guidance for management.

Case Report

A 55-year-old male presented with a 2-week history of progressive dyspnea, fatigue and hemoptysis. He had reportedly received medication for Brucella infection approximately 10 months ago [rifampin (Rifcap tablet, 300 mg, Koçak, Turkey) 300 mg po, 1 b.i.d. for 3 weeks plus doxycycline (Tetradox tablet 100 mg, Fako, Turkey) 200 mg po 1 daily for 6 weeks]. The diagnosis of brucellosis was made from a history of goat cheese ingestion, the symptoms and signs of fever, diaphoresis, appetite loss, headache, myalgia, low-back pain, raised level of Brucella agglutinin (1:160), and a positive bone marrow culture. Within 1 week of initiation of therapy, the patient became afebrile and the constitutional symptoms disappeared gradually. He remained well, the agglutinin concentration decreased, and the blood and bone marrow cultures were negative during the regular follow-up after discharge.

The patient had had falls and squatting ever since childhood and approximately 2 weeks prior to presentation the recent symptoms had appeared. Physical examination revealed a blood pressure of 110/75 mmHg, a heart rate of 120 beats/min and a body temperature of 38.1°C. Other findings were central cyanosis, pulmonary rales, clubbing, systolic murmur along the left sternal border and a third heart sound on the left precordium. Laboratory examina-

Fig. 1. Parasternal long axis view shows right ventricular hypertrophy, overriding aorta, and a wide membranous ventricular septal defect. RV, Right ventricle; LV, left ventricle; LA, left atrium; AO, aorta; IVS, interventricular septum.

Fig. 2. Apical 4-chamber view shows vegetation on the tricuspid valve. RV, Right ventricle; LV, left ventricle; LA, left atrium; RA, right atrium; TV, tricuspid valve.
tion showed a hemoglobin concentration of 11.2 g/dl, a white blood cell count of 6,800/mm³, a blood urea nitrogen of 129 mg/dl, a creatinine level of 3.7 mg/dl, a sedimen-
tation rate of 93 mm/h, a Po2 level of 49.9 mmHg, a PCO2:
level of 25.8 mmHg, and a systemic arterial O2 saturation
of 86.2%. An electrocardiogram demonstrated normal
sinus rhythm, incomplete right bundle branch block, and a
pathologically deviated right axis. Telecardiogram revealed
an enlarged aortic arch with an increased cardiothoracic
index. Transthoracic echocardiography with Doppler
revealed a membranous ventricular septal defect, enlarged
and overriding aorta, biventricular hypertrophy, a right
ventricular outflow tract gradient of 94 mmHg, and a left
ventricular ejection fraction of 63%. There were also a
grade 3 tricuspid insufficiency, and 2 separate masses
consistent with endocardial vegetation on the pulmonary
and tricuspid valves. The pulmonary and tricuspid masses
measured approximately 5×6 mm and 9×11 mm, respective-
ly (Figs 1,2). Bilateral multiple small subpleural nodules
with irregular margins indicating alveolar consolidation on
the apical segment of the lower lobe of the right lung were
detected with computed tomography (Fig 3). Abdominal
computed tomography showed splenomegaly and splenic
infection. Blood culture grew Enterococci colonies, which
were sensitive to vancomycin and gentamycin. After being
titrated to the dose/weight schedule, parenteral antibiotics
were given: vancomycin iv 500 mg/day (Vancocin-CP
flacon 500 mg, Lilly, Turkey) plus gentamicin iv 80 mg/day
(Genta ampul 40 mg, i.E. Ulugay, Turkey).

In the mean time, the dyspnea and fatigue had markedly
worsened and he had a weight loss of 12 kg. The patient
developed massive hemoptysis while on antibiotics and
required a blood transfusion of 10 units. He went into car-
diac arrest and did not respond to intensive resuscitation
performed during emergency bronchoscopic examination.

Discussion

This middle-aged patient had TOF complicated by
decompensated heart failure, infective endocarditis (IE)
and pulmonic involvement secondary to IE. Adults with
TOF are a special subset of patients with peculiar problems;
namely, the effects of prolonged cyanosis, polycythemia,
coeagulation defects, development of collateral vessels and
secondary myocardial dysfunction. Prolonged survival of
patients with uncorrected TOF is often associated with
a large bronchial to pulmonary collateral circulation and a
well-developed left ventricle.4–6 Chronic congestive heart
failure, secondary to the long-standing pressure overload
and consequent pathological changes in the right ventri-
cle,6,7 and arrhythmias8 are the most common causes of
death.

IE is another cause of death in adults with congenital
heart disease and streptococci are the most common or-
ganisms, followed closely by staphylococci.2,3 The present
patient had brucellosis, which was a differential diagnosis
for the IE. However, Brucella endocarditis was excluded
because the patient had an improvement in the clinical
signs, a decrease in the agglutinin concentration, and nega-
tive blood and bone-marrow cultures after therapy.
Endo-
coccus, which is the causative organism in 5–15% of
adult IE cases9 was isolated from blood cultures at last
admission. Enterococci are part of the normal gastrointesti-
nal flora and cause genitourinary tract infections. IE occurs
in older predominately male patients, with the urinary tract
as the likely portal of entry? In the present case we could
not determine the portal of entry, but we had limited time
or were not given permission for autopsy.

It is obvious that the IE significantly contributed to the
heart failure, continued embolism, and medically uncontrolled infection are among
the indications for surgical intervention for IE. In the
current era, late surgical repair of TOF is recommended
because the surgical risk is considered to be acceptable and
the long-term results have been gratifying in the rare series
reported; that is, short- and long-term mortality rates of
3–16% and 6–24%, respectively.10,11 The survival curve of
surgically treated patients in a study by Hu et al11 was
compared with age-matched medically treated patients in
the Danish data from 1949, as analysed by Bertranou et al,11
and the survival of the surgically treated group was found
to be significantly better than that of the medically treated
group. Unfortunately, those studies provide little informa-
tion to guide management decisions for a decompensated
patient. A preoperative functional class of IV improved
after surgery in some patients in some studies.12–13 and Hu et
al reported only one early postoperative death in a patient
with preoperative clinical class IV.11

The final cause of death in the present patient was mas-
Sive hemoptysis. It is known that, whereas the less severe
forms of hemoptysis occur most often in the presence of
pulmonary vascular obstructive disease or in patients with
an extensive bronchial collateral circulation or pulmonary
venous congestion14 massive hemoptysis almost always
represents rupture of a dilated artery in patients with con-
genital heart disease.15

In summary, this case provides a rare insight into the late
outcome of a middle-aged patient with uncorrected TOF.

References

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