Surgical repair of combination of Addison’s Disease and secundum type atrial septal defect and our postoperative follow-up principles

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INTRODUCTION

Atrial septal defect is a common congenital heart defect. Abnormal apoptosis and retarded developmental growth are proposed as pathogenic mechanisms[1]. Atrial septal defect is frequently reported with genetic syndromes. But, to the best of our knowledge, it has not been reported with autoimmune polyendocrine syndrome[2].

Adrenal insufficiency can be either primitive as the result of a destruction of the glands or secondary to a corticotropic failure. Adrenal insufficiency can appear as an acute event or in contrast arise progressively[3]. The chances of cardiovascular collapse leading to a diagnosis of Addison’s disease are rare. One report cited 3 of 60,000 cases in which this may have occurred[4].

CASE PRESENTATION

Our case was a 23-year-old female. She was suffering from dizziness, palpitation and easy fatiguability for 8 months. Investigations for this purpose revealed secundum type atrial septal defect and she was then referred to our clinic. Transthoracic echocardiography showed a 15 mm defect in the interatrial septum with a left-to-right shunt. Ejection fraction of this case was calculated as 65% and pulmonary arterial pressure was measured as 30 mm Hg. Particularly, at bicaval position, echocardiographic image of this ASD was optimal (Figure 1).

Superior rim of ASD was measured as 8mm whereas its aortic rim was measured as 3 mm. Therefore, it was found unsuitable to occlude this ASD via percutaneous transcatheter technique and surgical repair was recommended (Figure 2).
Cardiac catheterization pointed out no additional pathology. Qp/Qs was calculated 1.5.

Moreover, our case possessed Addison’s disease consistent with adrenal insufficiency diagnosed two months ago. For this purpose, he was taking 5 mg of oral prednisolone per day. She was consulted by Department of Endocrinology and perioperative recommendations and precautions were obtained.

She was operated under endotracheal general anesthesia and in supine position. As it was a part of our anesthesia and cardiopulmonary bypass protocol for its membrane stabilizing effect, also recommended by Endocrinology, 250 milligrams of intravenous methylprednisolone succinate was administered. Moreover, 50 milligrams of ranitidine was administered intravenously for gastric protection. Following a median sternotomy, pericardium was opened longitudinally. After heparinization, extra-corporeal circulation is established between the venae cavae and the ascending aorta. A cross clamp was placed on aorta and by antegrade intermittent isothermic blood cardioplegia from aortic root, cardiac arrest was established. Hypothermia was moderate (32ºc). A vent was placed via the right superior pulmonary vein. Standart right atriotomy was made. ASD was evaluated regarding its localization, size, other related cardiac structures and possible associated abnormalities (Figure 3).

We performed a primary closure of atrial septal defect (Figure 4).

Right atriotomy was closed in a standard fashion. Postoperative rhythm was sinus. She didn’t require inotropic support during weaning from cardiopulmonary bypass and early postoperative period. The post-operative course was uneventful with successful anatomical correction. The doses of corticosteroid treatment as recommended by Department of Endocrinology were given in Table 1. Moreover, gastric protection therapy with intravenous ranitidine HCl 2x50 mg per day was continued for 7 days.
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Figure 5

Table 1: Posology of the corticosteroid therapy administered postoperatively.

<table>
<thead>
<tr>
<th>Postoperative day</th>
<th>Corticotherapy dosage</th>
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<tbody>
<tr>
<td>1.</td>
<td>100mg methylprednisolone sodium succinate IV</td>
</tr>
<tr>
<td>2.</td>
<td>50mg methylprednisolone sodium succinate IV</td>
</tr>
<tr>
<td>3.</td>
<td>50mg methylprednisolone sodium succinate IV</td>
</tr>
<tr>
<td>4.</td>
<td>50mg methylprednisolone sodium succinate IV</td>
</tr>
<tr>
<td>5.</td>
<td>5+2.5 mg per day prednisone P.O.</td>
</tr>
<tr>
<td>6.</td>
<td>5+2.5 mg per day prednisone P.O.</td>
</tr>
<tr>
<td>7.</td>
<td>5+2.5 mg per day prednisone P.O.</td>
</tr>
<tr>
<td>8. and everyday</td>
<td>5+2.5 mg per day prednisone P.O.</td>
</tr>
</tbody>
</table>

Postoperatively an echocardiographic investigation was revealed no residual shunt for the repaired ASD. She was followed at our outpatient and endocrinology outpatient clinic without additional problem.

DISCUSSION

Congenital heart defects are the most common birth defects and represent an increasing proportion of adolescent. While many of these patients have undergone successful palliative or corrective surgery with excellent functional result[s].

In the human heart, septation occurs between 4 and 7 weeks of development. Cardiac looping and chamber formation bring the contributing structures into position to engage in septation[s]. Septation defects and patent ductus arteriosus are the most common human cardiovascular malformations (CVMs). Genetic factors play a major part in the origin of these malformations[s].

The protein EP300 and its paralog CREBBP (CREB-binding protein) are ubiquitously expressed transcriptional co-activators and histone acetyl transferases. The gene EP300 is essential for normal cardiac and neural development, whereas CREBBP is essential for neurulation, hematopoietic differentiation, angiogenesis and skeletal and cardiac development. The CBP/p300-interacting transactivator with ED-rich tail 2 (CITED2) binds EP300 and CREBBP with high affinity and regulates gene transcription[s]. The study of Bamforth et al. show that Cited2-/- embryos die with cardiac malformations, adrenal agenesis, abnormal cranial ganglia and exencephaly. The cardiac defects include atrial and ventricular septal defects, overriding aorta, double-outlet right ventricle, persistent truncus arteriosus and right-sided aortic arches[s].

Assessment of the patient with known or suspected congenital heart defects requires a careful history, physical examination, and noninvasive assessment[s].

Echocardiography was the most frequently used investigative modality in all defect sizes and types[s]. The transthoracic approach was successful in capturing sufficient data to create 3-D images, which can provide an accurate assessment of secundum ASD[s]. Surgical closure of ASD has a low perioperative mortality and morbidity[s].

The treatment of adrenal insufficiency includes substitutive doses of mineralo and/or glucocorticoids and, as often as possible, etiologic therapy. For this reason, as soon as the diagnosis of adrenal insufficiency has been made, a main point is to determine the cause of the endocrine failure[s].

References

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