may aid in management of these patients. The treatment of TAPE must address the immune response targeting the nervous system and the underlying thymoma.

Thirty-three percent of patients with TAPE died, all as a consequence of the nervous system compromise. The majority of these patients who died had stage I thymomas. Early Masaoka stage thymomas may produce more functional and neurotoxic autoantibodies than their malignant counterparts.⁴ Therefore, outcome is driven more by ongoing neurologic insult than the underlying thymic disease. Only 21% of patients with TAPE achieved symptom-free survival; all of these patients had complete resection of their thymoma.

With TAPE, it is incumbent on the clinician to make the link between thymoma and encephalitis before progressive neurologic symptoms and death ensue. The reported cases

suggest a role of antineuronal autoantibody production in the pathogenesis and, therefore, in the rationale for treatment options. We believe that immunotherapy and complete resection confer the best chance of symptom-free survival.

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Total correction of tetralogy of Fallot associated with rhabdomyoma in the right ventricle

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Primary cardiac tumors are a rare disease, with a low incidence. Rhabdomyoma, a type of benign primary cardiac tumor, is the most common primary cardiac neoplasm in children and occurs almost exclusively in children. We reported a rare case of an infant with tetralogy of Fallot associated with rhabdomyoma in the right ventricle.

CLINICAL SUMMARY

An 8-month-old male infant had cyanosis and a heart murmur at birth. Physical examination revealed cyanosis, a systolic murmur at the second and third intercostal spaces left of the sternum, and a weakened pulmonary second heart sound. Transthoracic echocardiography showed typical anatomic appearance of tetralogy of Fallot and a 2×2 -cm mass in the right ventricular cavity. Multiple-slice com-

puted tomography confirmed the findings from echocardiography and revealed that the mass was likely to attach to the right side of the interventricular septum (Figure 1). An on-pump open cardiac operation was performed. Cardiopulmonary bypass was established by cannulation of the aorta and venae cavae, and systemic temperature was kept around 28°C for the intracardiac procedure. Cold blood cardioplegia was used to induce and maintain cardiac arrest. A longitudinal incision was made crossing the pulmonary valve annulus, and the $2 \times 2 \times 1$ -cm mass was found attached to the right side of interventricular septum close to cardiac apex, surrounded by muscular trabeculae (Figure 2, A). The base of the mass was about 1.5×1 -cm wide. This mass had a color similar to normal myocardium with a smooth surface and irregular shape (Figure 2, B). The cardiac mass was resected completely, with care taken to cause as little injury to the normal myocardium as possible, followed by correction of tetralogy of Fallot, including infundibulectomy, repair of ventricular septal defect, and transannular pericardial patch (Figure 2, C). This infant was successfully weaned from cardiopulmonary bypass. The mass was investigated pathologically. The immunohistochemical assays showed that stainings of actin (Figure 2, D), desmin, and vimentin were strongly positive. However, staining of S-100 was negative. These histologic findings proved that this cardiac mass was a rhabdomyoma. During

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FIGURE 1. Multiple-slice computed tomography revealed a 2- × 2-cm mass in the right ventricular cavity, probably attaching to the right side of interventricular septum.

the first 2 days after the operation, the infant had low cardiac output syndrome. Positive inotropic agents including dopamine, epinephrine, milrinone, and deslanoside (Cedilanid) were administered. On the ninth day, the infant was weaned from mechanical ventilation with stable cardiac hemodynamics and vital signs.

DISCUSSION

Primary cardiac tumors are rare, with an incidence of 0.001% to 0.28% by autopsy. In this case, the cardiac mass was confirmed to be rhabdomyoma of the right ventricle,

which is the most common primary cardiac neoplasm in children, occurring in the majority before 1 year of age.¹ Rhabdomyomas are usually multiple and intraventricular.² However, this one was solitary and attached to the right side of the interventricular septum close to apex. In this case, transthoracic echocardiography detected the congenital heart anomaly associated with the cardiac mass, which has a detection rate of 95.2% for the diagnosis of intracardiac tumors.¹ Multiple-slice computed tomography provided valuable information about the existence, size, position, and attachment mode of the mass in this case, which helped us greatly in

FIGURE 2. A, During the operation, a mass was found attaching to the right side of interventricular septum close to the cardiac apex, surrounded by muscular trabeculae. B, The mass had a smooth surface and irregular shape. C, The heart with resection of the cardiac mass and correction of tetralogy of Fallot. D, Immunostaining of actin was strongly positive for the cardiac tumor.

working out a strategy of surgery. When possible, resection with cardiopulmonary bypass is the standard surgical procedure for benign primary cardiac tumors and can lead to excellent outcomes with low recurrence rates.^{3,4} Tetralogy of Fallot occurring with primary cardiac tumor is very uncommon. This complex type of cardiac anomaly made the operation more difficult in this case. From this case, we found that minimizing the injury to normal myocardium was essential for the maintenance of postoperative cardiac function when the tumor was removed, because postoperative low cardiac output syndrome occurs frequently in children with tetralogy of Fallot. After the operation, positive inotropic agents should be administered actively and prolonged mechanical ventilation is also necessary. In this case, the infant had low cardiac output syndrome during the first 2 days after the operation. After

receiving appropriate therapy, he was weaned from mechanical ventilation on postoperative day 9 with stable cardiac hemodynamics and vital signs. Successful surgical treatment of this rare case may provide some valuable experience for the complex type of primary cardiac tumors associated congenital heart disease such as tetralogy of Fallot.

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