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Wilms Tumor with Intracardiac Neoplastic Extension

Key Words

Wilms tumor
Nephroblastoma
Intracardiac tumor thrombus
Bypass, cardiopulmonary

Abstract

A rare case of a Wilms tumor with intracardiac neoplastic extension is presented. The good prognosis of even extensive nephroblastoma is the reason for attempting a radical surgical approach to patients with intracardiac tumor thrombus. The exact preoperative delineation of the intracardiac tumor extension is essential for a safe and successful operation. Ultrasonography in combination with echocardiography is the preferred modality for detecting intracardiac extension.

Introduction

Nephroblastoma (Wilms tumor) is one of the most common intra-abdominal tumors of childhood [1]. It grows by direct extension [2] and exhibits a marked tendency to invade vascular structures. A rare but dangerous complication is the extension of the nephroblastoma into the right atrium via the inferior vena cava (IVC). Caval tumor thrombosis occurred in 4.1% of 1,865 patients treated within the National Wilms Tumor Study 3; in 0.9% of the patients, this thrombus extended into the right atrium [3]. This extension may cause complications such as right heart inflow/outflow obstruction and pulmonary embolism. Due to modern combined therapeutic regimes, such as chemoradiotherapy and surgery, children suffering from nephroblastoma can be expected to have disease-free survival rates of between 50 and more than 90% depending on stage and histology [4]. This good prognosis of even extensive nephroblastoma is the reason for attempting a radical surgical approach in patients with intracardiac tumor thrombus. The level of vena caval involvement alone does not seem to be of prognostic sig-

nificance [5]. Thus radical surgical excision of the tumor is recommended followed by chemotherapy. We present a case of a patient with nephroblastoma and intracardiac extension (ICE).

Case Report

In 1987, a 4.5-year-old girl was referred to our institution because of recurrent febrile episodes with abdominal pain of 4 weeks duration. Physical examination revealed a large upper left abdominal tumor. Physical ultrasound showed an inhomogenous partially cystic mass (10 × 8 × 7 cm) in the region of the left kidney, multiple masses of up to 2 cm in diameter on both sides of the abdominal aorta, and an almost complete occlusion of the IVC combined with a mass in the right atrium. The parenchyma of the liver appeared to be normal, the hepatic veins seemed to be congested. Intravenous pyelography showed no visualization of the left kidney 2 h after injection of contrast media. Distal cavography demonstrated obstruction of caval flow at the level of D 12 with collateral drainage via lumbar veins. Multiple pulmonary metastases were detected by X ray; skeletal or bone marrow metastases were ruled out by ⁹⁹Tc-diphosphonate scintigraphy and bone marrow cytology. Laboratory investigations were normal except for an elevated serum LDH (707 U/l). The presumptive diagnosis was Wilms tumor of the left kidney with tumor

thrombosis in the IVC/right atrium and multiple pulmonary metastases (stage IV).

First, left tumor nephrectomy and excision of all para-aortal lymph nodes was performed. Subsequently, the tumor thrombus in the IVC and right atrium was removed under the conditions of a cardiopulmonary bypass and a short period (10 min) of deep hypothermic circulatory arrest. The postoperative course was uneventful.

Histopathological examination revealed a nephroblastoma with blastemal predominance. Five courses of a 9-week regimen including vincristine and pulses of cyclophosphamide, Adriamycin and actinomycin D were given. The primary tumor region was irradiated with 19.2 Gy, both lungs received 12.0 Gy. The duration of postoperative treatment was 1 year. The pulmonary metastases regressed completely. At present, 4 years after the end of treatment, the girl is relapse-free and growing normally.

Discussion

The prerequisite for a well-planned, safe and successful operation is the exact delineation of the intravasal tumor extension [6]. This has been greatly facilitated by the newer techniques of noninvasive imaging. The preferred modality is ultrasonography [7] in combination with echocardiography [8]. Especially two-dimensional echocardiography with Doppler analysis and color flow mapping is presently considered as the most useful and reliable method for detecting ICE [9]. The computerized axial tomography scan seems to be of less diagnostic value [3]. The nuclear magnetic resonance scan as a new promising non-invasive imaging technique needs further experience [10]. In rare cases with inconclusive diagnostic results or specific questions, the invasive method of inferior cavography and/or brachial right heart catheterization is justified.

With angiography of the IVC, as the most sensitive tool, the correct diagnosis of intracaval thrombus is achieved in nearly 100% [3]. Intracaval tumor extension below the level of the hepatic veins does not imply a particularly difficult surgical procedure. However, as the intravasal tumor extends to the diaphragm or right atrium and beyond, the hazards and difficulties of complete resection increase significantly [11]. Therefore, in the 1980s the use of the combined techniques of cardiopulmonary bypass and deep hypothermic circulatory arrest was introduced to assist in the resection of tumors that extend into the IVC above the level of the hepatic veins [6]. This approach enables the surgeon to operate in a totally bloodless field. Hypothermic arrest within given time limits is a safe and well-accepted procedure, especially in surgery for congenital heart disease [12].

Long-term survival of children with Wilms tumor and ICE is stage-dependent (2-year relapse-free survival in 78% in stage III, 72% in stage IV) [5]. These encouraging results can be achieved by surgery followed by a combined therapy with radiation and multiagent chemotherapy [4]. On the other hand, primary chemotherapy was shown to lead to a complete remission of the tumor in about 75% [3]. However, the extension of the tumor beyond the diaphragm confers particular risks, namely tricuspid valve obstruction [1], pulmonary embolism, tumor relapse or incomplete response to chemotherapy as illustrated by case reports in the literature [3].

Therefore, an aggressive surgical approach using the safe facilities of a cardiopulmonary bypass and hypothermic arrest is advocated in children with ICE reserving primary chemotherapy alone for unresectable tumors.

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