Preoperative giant sacrococcygeal teratoma embolization in a newborn – A case report and a review

J. Guitart, M. Teixidor, N. Brun, S. López, E. Criado, N. Romero

Parc Taulí Healthcare and University Corporation. Sabadell, Barcelona (Spain).

Abstract

Sacrococcygeal teratoma (SCT) is the most frequent congenital germ cell tumor. Patients have a higher risk of perinatal complications and death, with bleeding and cardiac decompensation being the most common causes of neonatal mortality.

This is the case of a 35-week preterm newborn with a large SCT diagnosed at ultrasound screening in the second trimester. Preoperative selective embolization of the middle sacral artery and total surgical resection were performed postnatally with minimal blood loss. The patient was discharged at 25 days of life with a normal physical examination.

Selective embolization prior to giant SCT resection is feasible and appears as a safe and useful technique in the control of perioperative bleeding.

Key Words: Sacrococcygeal teratoma; Preterm newborn; Embolization; Middle sacral artery; Preoperative embolization.

Embolización preoperatoria de teratoma sacrococcígeo gigante en un recién nacido. Reporte de un caso y revisión

Resumen

El teratoma sacrococcígeo (TSC) es el tumor congénito de células germinales más frecuente. Los pacientes afectados tienen un mayor riesgo de complicaciones perinatales y muerte, siendo la hemorragia y la descompensación cardiaca las causas más comunes de mortalidad neonatal.

Presentamos el caso de un recién nacido pretérmino de 35 semanas con un TSC de gran tamaño diagnosticado por ecografía en el segundo trimestre. La embolización selectiva preoperatoria de la arteria sacra media y la resección quirúrgica total postnatal se realizaron con una mínima pérdida de sangre. El paciente fue dado de alta a los 25 días de vida con un examen físico normal.

La embolización selectiva antes de la cirugía de resección del TSC gigante es factible y aparece como una técnica segura y útil en el control del sangrado perioperatorio.

Palabras Clave: Teratoma sacrococcígeo; Recién nacido prematuro; Embolización; Arteria sacra media; Embolización prequirúrgica.

INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most frequent germ cell tumor in pediatric age and the most common perceptible fetal neoplasia[1]. Typically, it is diagnosed at prenatal control ultrasound screening as an intrauterine mass. Lesions over 10 cm are associated with high perinatal mortality, especially if they are hypervascular, if they have been caused by intraoperative bleeding, or if they are complications arising from congestive cardiac insufficiency. These tumors are usually irrigated by a hypertrophic middle sacral artery, so arterial ligation or control are a key step in the surgical procedure. A comprehensive literature search showed three preoperative SCT embolization cases only[2-4]. This is the case of a patient born at 35 weeks gestation with type 1 giant SCT successfully embolized prior to surgical removal. The objective of this article was to describe the endovascular procedure and discuss benefits and risks.

CINICAL CASE

Preterm 35-week newborn diagnosed with giant SCT prenatally. Ultrasound screening in the second trimester and subsequent fetal MRI at 29 weeks gestation confirmed the presence of a type 1 SCT according to Altman's classification. No signs of cardiac insufficiency were noted at subsequent fetal echocardiograms. An urgent cesarean section was carried out as a result of a premature rupture of the membranes and anomalies in the associated cardio-

Corresponding author: Dr. Josep Guitart Giménez. Parc del Taulí, 1. 08208 Sabadell, Barcelona (Spain).
E-mail address: jguitartg@gmail.com

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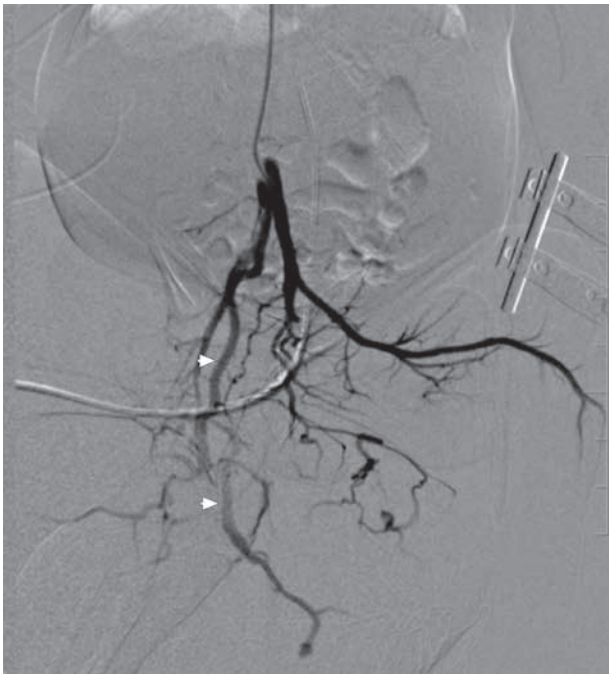

Figure 1. Middle sacral artery hypertrophy (arrow tips) with significant neovascularization in the tumor bed.

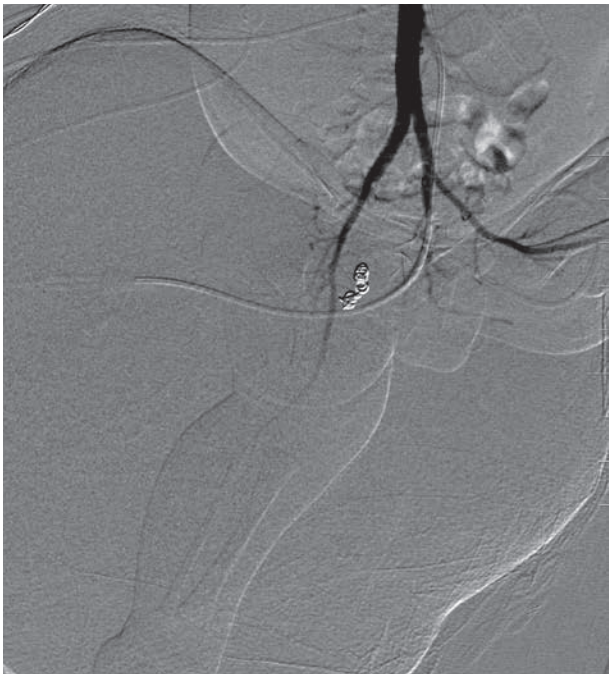

Figure 2. Aortoiliac angiogram following embolization with Gelfoam® and microcoils.

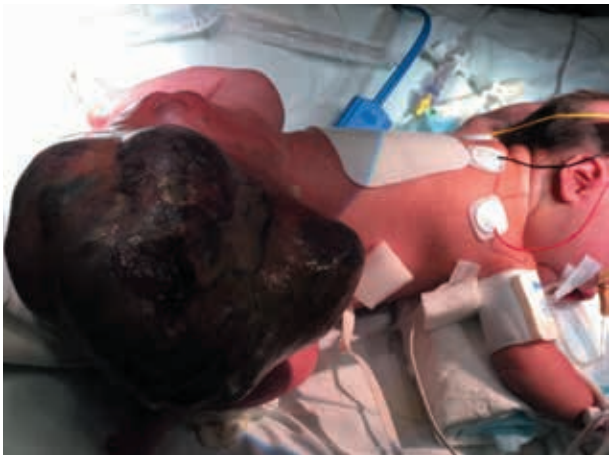

Figure 3. Tumor appearance prior to the procedure (18 hours following embolization) showing signs of ischemia and necrosis.

tocographic record. Patient weight at birth, including the tumor, was 4,030 g. SCT was approximately 19 x 15 cm in size and had ulcerations and bleeding signs on its surface.

An echocardiogram was performed at 24 hours of life. It demonstrated a 3 mm persistent ductus arteriousus with left-right communication – which was normal considering the hours since birth – and no further relevant disorders (73% ejection fraction).

Owing to ulcerations and size, elective surgical removal at 24 hours was indicated. In order to control tumor irrigation through the middle sacral artery, decision was made to try preoperative embolization under general anesthesia 18 hours following delivery.

Given that the common femoral artery approach was not feasible due to the position of the patient's limbs – as a result of the tumor's size –, the left common carotid artery (CCA) was used following surgical exposure. An abdominal aortography was performed by means of a manual contrast injection using a 4F vascular introducer sheath and a 4F multipurpose catheter. 250 IU of heparin were administered through the introducer sheath. Lower abdominal aorta angiogram demonstrated the presence of a large hypervascularized mass irrigated by a significantly hypertrophic middle sacral artery (Fig. 1) and, to a lesser extent, by some lateral sacral branches of the left hypogastric artery. The middle sacral artery was selectively catheterized using a 2.7F microcatheter (Progreat®, Terumo Europe) and successfully embolized with Gelfoam® (Pfizer, Spain) and two proximal pushable microcoils (Fig. 2). Embolization was performed with 5 ml of iodinated contrast diluted in 10 ml of saline solution. Total procedure time, including the surgical dissection of the left CCA, was 90 minutes. The patient was moved to the neonatal ICU without further complications. The elective procedure was carried out the following day, at 38 hours of life, through a posterior sacral access, with a minor intraoperative blood loss that required a 5 ml/kg red blood cell concentrate transfusion, and with a 9.8 g/dl final hemoglobin level. Prior to removal, the tumor had a purplish ischemic appearance with necrotic areas (Fig. 3).

The tumor's pathological anatomy confirmed diagnosis of immature congenital teratoma (with a maximum diameter of 18 cm) and fragment of coccyx with adjacent teratomatous elements (pancreatic tissue). The patient was discharged at 25 days of life with a normal physical examination.

DISCUSSION

This is the case of a successful preoperative SCT embolization using a left common carotid artery (CCA) approach. CCA access, either through arteriotomy, or more recently, through percutaneous access[5], has already been described in infants with congenital cardiopathy undergoing cardiac catheterization. However, there is little information on its use for other vascular procedures. To our knowledge, there are only 3 SCT cases published where preoperative hemostatic control was carried out using an endovascular approach, and only 1[3] where the CCA approach was used.

Prenatally diagnosed SCT is difficult to manage as it is highly unpredictable. Perinatal mortality ranges from 40% to 50%[6].

Imaging characteristics, particularly >10 cm tumor volume, and associated with solid component, can provide prognostic information[6]. Other factors associated with high risk SCT include polyhydramnios and rapid tumor growth. Fetal cardiac dysfunction (often associated with non-immune hydrops), spontaneous tumor bleeding, and preterm delivery are also highly associated with perinatal mortality and morbidity[7].

The objective of intrauterine treatment measures is to limit the cardiovascular impact of the tumor mass, as well as to allow for normal fetal growth and maturation. In the presence of selected high risk SCT factors, fetal surgery can be indicated.

Prenatal open fetal surgery is recommended in high risk fetuses and in case of hydrops when it appears at a gestational age too early for delivery and neonatal care (typically between weeks 28 and 32)[8].

Pregnancy termination is another option for parents. Low risk SCT patients are usually born through cesarean section after 36 weeks of gestation. High output cardiac insufficiency, internal tumor bleeding, and perioperative bleeding are the most frequent premature death causes and are highly associated with tumor size[9].

Definitive treatment involves performing a complete tumor resection, which typically includes coccyx removal. Early tumor resection is associated with a better prognosis. The surgical approach depends on tumor size and pelvic structure infiltration. Surgical resection can be complex, with a high risk of uncontrollable tumor bleeding and hemodynamic instability, which can be life-threatening. Mortality in SCT newborns as a result of bleeding represents nearly 70% of general mortality in the neonatal period. To prevent this risk, an adequate surgical dissection should be carried out. Early occlusion of the arteries irrigating the tumor can also prove useful.

The middle sacral artery usually irrigates the SCT, and can be as hypertrophic as the common iliac artery, which may cause vascular steal syndrome[10]. The tumor can also be irrigated by both internal iliac artery branches, primarily through the lateral sacral arteries.

There are few cases published in the literature describing tumor devascularization prior to tumor removal by ligating the middle sacral artery or the internal iliac arteries, or by performing a temporal clamping of the abdominal aorta[11].

To our knowledge, there are only 3 cases published where preoperative hemostatic control was carried out using an endovascular approach.

In 2006, Cowles et al. published the case of a 36-week newborn with a large type 1 SCT undergoing arterial embolization and subsequent direct radiofrequency percutaneous ablation for hemostatic control purposes prior to surgical resection. In this patient, the vascular access was carried out through the right common femoral artery. The middle sacral artery, the lateral sacral arteries, and the gluteal artery were embolized using Gelfoam® and acrylic glue[2].

In 2011 and 2013, respectively, Lahdes-Vasama et al.[3] and Rossi et al.[4] described the preoperative devascularization of a giant SCT using an endovascular approach only. In the first case, the patient was a 30-week preterm newborn; and similarly to our case, common carotid artery access was decided upon.

The position of the lower limbs in semi-abduction owing to the tumor's size made us rule out the femoral approach. However, a more favorable catheterization of the middle sacral artery was achieved using a supra-aortic approach. In our short experience, an adequate selection of the arterial access is crucial, since this is the most demanding and most important step in terms of procedure time. Therefore, it should be anticipated and discussed with the surgical team. The small size of arterial access, the higher propensity to arterial spasm it has, and the fact that femoral access can be unfeasible as a result of pelvic mass characteristics are the main causes of difficulty in arterial access.

Liquid management in the preterm newborn is key as it limits the amount of liquids that can be injected during the angiogram and the embolization. This is another important technical point.

Regarding complications, apart from those secondary to puncture (hematoma, pseudoaneurysm, etc.) and catheterization (dissection, perforation, thrombosis, embolism, etc.), there are other complications specific to the anatomical site under embolization.

Postembolization syndrome, which has been profusely described in hepatic chemoembolization and fibroid embolization, is not a complication but one of the most frequent

side effects of arterial embolization. It presents with pain, fever, and nausea/vomit, typically mild and self-limited within the first 72 hours.

Cutaneous, bladder, and intra-abdominal organ necrosis has been described in embolizations in the pelvic area, as well as neurological damage (paraparesis, neuropathy) and even medullary ischemia, but with a low incidence[12].

In our case, as well as in the other cases published in the literature, there were no complications related to the technique or the area embolized. Elective surgery immediately after arterial embolization was technically successful and no significant blood loss was noted.

To sum up, selective embolization of the middle sacral artery prior to giant SCT surgical resection is feasible and appears as a safe and useful technique to control perioperative bleeding and reduce neonatal death risk.

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