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CASE REPORT

Congenital immature teratoma in infant

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ABSTRACT

Teratomas are rare neoplasms. They account for 3% of all childhood tumors; 40–70% of them are located in the sacrococcygeal and presacral regions. Head and neck locations of the tumor are uncommon, account for only 2%. This article presents a case of a Polish Caucasian male infant born at 30 weeks of gestation, from the 3rd pregnancy, the first delivery by c-section. The infant's general condition was very severe. Birth weight was 2980 g. A large craniofacial tumor was prenatally detected in the newborn, with the suspicion of immature teratoma, which was later confirmed in postnatal examinations. The infant was operated on the 2nd day of life and the tumor size was 25–30 cm. After the surgery, birth weight decreased to 1600 g. Imaging tests provides prenatal diagnosis of craniofacial teratomas, which enables to plan early surgical treatment of the newborn with appropriate postoperative care, which increases the chances of survival.

KEY WORDS:

newborn, congenital neoplasm, craniofacial teratoma, teratoma, giant head and neck mass.

INTRODUCTION

Congenital teratomas are rare neoplasms originating from three germ layers and showing a different histological structure than the tissues characteristic for a given anatomical region [1]. They account for 3% of all childhood tumors; 40–70% of them are located in the sacrococcygeal and presacral regions. Head and neck locations of the tumor are uncommon, account for only 2% [2]. Depending on the size and location of the tumor, they can cause a variety of symptoms, including breathing and eating difficulties. In this article, we present the case of a newborn who was surgically treated for a congenital, immature craniofacial teratoma.

CASE REPORT

A male newborn from the third pregnancy, the first delivery, was born in the 30th week of gestation by caesarean section. Prenatal magnetic resonance imaging (MRI) of the fetus revealed a cystic-solid tumor of the right side of craniofacial region, which destroyed the subtemporal fossa together with the soft tissues of the face, reaching the midline – Teratoma immaturum was suspected. The course of pregnancy was complicated by premature rupture of the membranes for 7 days. The mother received a course of betamethasone steroid therapy in the 1 × 12 mg regimen for two days (total dose 24 mg).

Due to the tumor, after the fetal head was delivered, the baby assumed a forced position with the face facing the mother's left thigh. Furthermore the mother started to extensively bleed from the uterus excluding the possibility to keep prolonged placental circulation. Both factors precluded the EXIT procedure to be performed. Thus, immediately after birth, the umbilical cord was cut. The birth weight of the child with the tumor was 2980 g, the head circumference was 29 cm (84 percentile), and the body length was 39 cm (47 percentile). On the right side of the

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FIGURE 1. The photo of the patient before the surgery on the first day of life

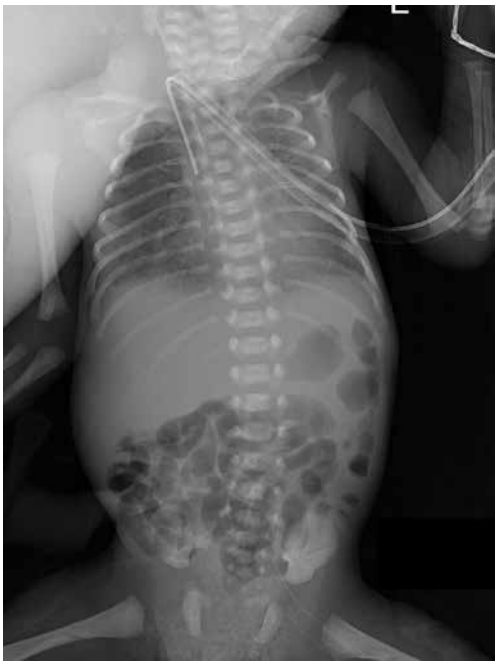

FIGURE 2. Chest X-ray with visible signs of respiratory distress syndrome I/II degree

face and neck, a large tumor was found, about 25–30 cm in diameter. The tumor was fluctuant, covered with skin with dilated blood vessels and petechiae (Figure 1).

After birth, breathing disorders were found. Due to the pressure of the tumor mass on the anatomical structures of the head and neck, attempts at endotracheal intubation were unsuccessful. Mechanical ventilation was applied with NeoPuff using a face mask. The child required a heart massage and dosage of adrenaline (total supply 4 × 0.3 ml of adrenaline 1 : 10000). An emergency tracheostomy was performed, then conventional mechanical ventilation was used. Because of the severe general condition of the newborn, a surfactant was administered intratracheally at a dose of 240 mg (200 mg/kg estimated body weight without tumor). Clinical improvement was achieved and saturation increased from 58–48–55% to 95–98%.

After the child was transferred to the Neonatal Intensive Care Unit, conventional mechanical ventilation was continued. Chest X-ray revealed respiratory distress syndrome I/II degree (Figure 2).

Due to circulatory instability, caused by extracellular volume shift to the tumor, the vascular bed was filled with crystalloids and colloids. The child required a supply of catecholamines (dobutamine 20 mcg/kg/min, dopamine 10–20 mcg/kg/min, adrenaline 0.16–0.3 mcg/kg/min and noradrenaline 0.7 mcg/kg/min). The patient's diuresis was supported with Furosemide. Coagulation disorders were treated with administration of multiple preparations of fresh-frozen plasma with pathogen reduction (15 ml/kg), cryoprecipitate (10 ml) and a concentrated irradiated filtered platelets (15 ml/kg). The patient was anemic therefore he received a concentrate of filtered irradiated red blood cells (at a dose of 15 ml/kg). Due to the patient's serious condition, the initial administration was total parenteral nutrition through the central venous catheter.

The features of sub/epidural haemorrhage and 2–3 mm of fluid spaces located near the right temporal and parietal lobe were described in the trans-ophthalmic ultrasound, performed on day 1 of life. In addition, the features of the immaturity of the brain tissue. The α-fetoprotein (AFP) was measured to be 130 000 ng/ml, while β-human chorionic gonadotropin (β-hCG) was normal.

On day 2 of the child's life, diagnostic imaging of the tumor was performed. In the computed tomography (angio computed tomography – CT) scan of the head and neck (Figure 3, 4), a cystic-solid tumor was described. It was located on the right side of the craniofacial region and neck with dimensions of approx. 210 × 200 × 80 mm. The tumor consisted mostly of a large cystic part and contained a richly vascularized solid element measuring approximately 50 × 31 mm. The study showed that the solid part of the tumor, containing few calcifications, reached the midline and adjoined the mandible and sub-temporal fossa. Its vascularization came from the external carotid artery.

On day 3 of the patient's life, the tumor was resected under general anaesthesia. During the procedure, the water-clear fluid was aspirated from the lumen of the cyst and the solid part was resected. The right external carotid artery was ligated. The tumor consisted of a fragment of the sidewall of the pharynx, the defect of which was covered with the patient's own tissues after resection. The right facial nerve was damaged intraoperatively, which was the cause of its peripheral paralysis and right eyelid insufficiency. Redundant skin tissue was removed (Figure 5).

The histopathological examination of the lesion revealed a tumor with immature neuroepithelial weaving

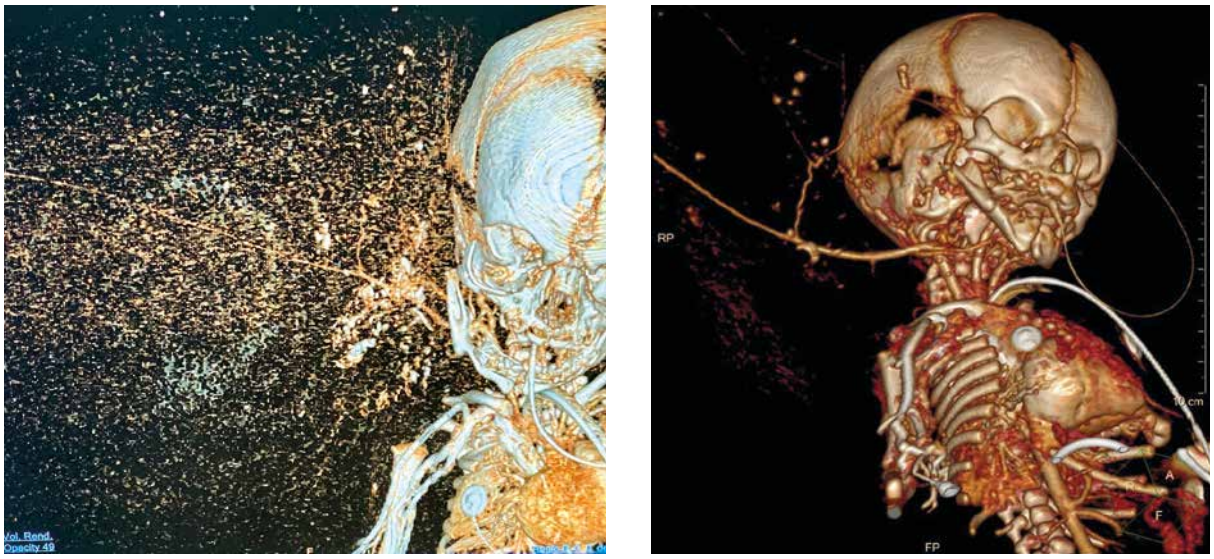

FIGURE 3, 4. Computed tomography examination of blood vessels (angio computed tomography) of the head and neck of a newborn with a visible cystic-solid tumor on the right craniofacial side and neck, dimensions approx., 210 × 200 × 80 mm

containing glandular and lymphatic structures and salivary gland tissue. This image corresponded to Teratoma immaturum. After tumor resection, the patient's body weight was significantly reduced and amounted to 1600 g.

Tumor resection contributed to the gradual stabilization of the child's condition, which made it possible to reduce the doses of catecholamines and eventually to discontinue Adrenaline and Noradrenaline on the 8th day of life, and dopamine and dobutamine on the 16th day of life.

On the 6th day of the newborn's life, a CT of the head was performed, which showed extensive cerebral bleeding above the left parietal and occipital lobe, along the crescent of the brain and the cerebellar tent, and in the ventricular system. The median structures were moved approx. 1 cm to the right side. The patient was under neurosurgical care. Due to the gradual, natural evolution of changes, neurosurgical treatment was abandoned.

After stabilization of the general condition, from the 7th day of life, trials of trophic feeding through a gastric tube were started. On the next day, due to difficulties with food supply, an abdominal scan was taken and a seriogram of the upper gastrointestinal tract was performed, which confirmed the perforation of the oesophagus. As a result, a right-sided pneumothorax developed, which was decompressed with the active drainage of the pleura. The newborn was given a gastric splint and was not fed enterally.

From the 23rd day of life, enteral feeding with mother's food was reintroduced in increasing amounts with good tolerance.

The patient was under the constant care of a neurologist. On the 31st day of the child's life, amplitude integrated electroencephalography (aEEG) test was performed, which showed short inserts of the record with reduced amplitude and a rather poorly marked variability of sleep and wakefulness. There was no recorded pattern characteristic of neonatal seizures, which were also not observed clinically.

FIGURE 5. The photo of the patient after the surgery

Due to the persistent respiratory failure, on the 35th day of life, a 7-day course of betamethasone steroid therapy was applied, thanks to which the demand for oxygen with the oxygen content in the respiratory mixture (FiO2) 0.6–0.35 was reduced. The patient was mechanically ventilated until the 38th day of life, then non-invasive respiratory support (continuous positive airway pressure) was used.

At 3 months of age, the patient had seizures and aEEG showed seizure changes. For this reason, anticonvulsant treatment (levetiracetam) was initiated. In the control EEG, performed at the age of 3.5 months, no seizure changes were found, therefore, it was decided to gradually discontinue the antiepileptic drugs. A follow-up MRI examination was performed. Extensive malacic lesions were found, which covered the entire left parietal and

partially occipital lobe on the left side, with the formation of a painting cavity connecting with the occipital horn and temporal horn of the left lateral ventricle. The child did not require any neurosurgical intervention.

The patient was diagnosed with Retinopathy of prematurity therefore, an intraocular Lucentis injection was performed (1.15 mg in each eye).

A neurological examination performed at 4 months of age (8 weeks of corrected age) showed poor motor skills with a flexion-extension pattern and movements of constant speed and amplitude. The infant had variable tension with a tendency to increase in the lower limbs. Thumb was placed in abduction. Deep reflexes were present, while the tension in the head-torso axis was reduced. Lacking the ability to coordinate sucking and swallowing gastrostomy was performed.

The patient was transferred to a center preparing for home respiratory ventilation. He is under multidisciplinary care, with regular examinations in the ENT, neurology and ophthalmology clinics. The child requires constant physiotherapy, rehabilitation and visits to a neurologist. It is also under constant oncological observation, with periodic inspections of tumor markers: AFP and β-hCG which stayed within normal limits.

Right now, the patient is 2.5 years old. He is sitting by himself, crawling and sometimes standing in orthoses. The child doesn't speak. He still has a tracheostomy and a percutaneous endoscopic gastrostomy. He can drink and eat pulp, still learns to bite. The patient enjoys playing with his siblings. He follows toys with his eyes. He is under the care of a speech therapist and physiotherapist.

DISCUSSION

Congenital head and neck tumors and malformations may cause direct postnatal upper airway obstruction, asphyxia and death. With the introduction of ultrasound screening programs, congenital head and neck pathology can frequently be diagnosed prenatally and the scenario of postnatal airway obstruction can be prevented.

Depending on the origin, obstructive head and neck lesions are classified as extrinsic or intrinsic to the fetal airway. Among extrinsic lesions, head and neck teratomas are the most common cause of fetal airway obstruction.

Many studies have found a high rate of perinatal mortality caused by tumor obstruction of the airways. In these patients, the EXIT procedure can be used, which involves opening the airways during cesarean delivery, before clipping the umbilical cord, which enables the maintenance of blood flow between the placenta and the fetus and reduces the risk of prolonged hypoxia [3]. In the described case, it was not possible to use the EXIT procedure, because the significant size of the tumor caused the fetus to be positioned in such a way that it prevented the rotation of the head to protect the respiratory tract and maternal bleeding therefore it was decided to detach earlier.

In patients with no evidence of airway obstruction during labor, tracheal intubation is still recommended due to the risk of tumor bleeding and consequent airway obstruction.

Despite the fact that teratomas are a rare group of childhood neoplasms, with a frequency of 3%, they are the most common neoplasms of the neonatal period [4]. These tumors are usually benign and come from the ectoderm, endoderm, and embryonic mesoderm. The mechanism of the formation of teratomas is still not fully understood. The most popular hypothesis assumes the occurrence of abnormalities during the migration of totipotent cells from the yolk sac during embryogenesis [5].

Although teratomas can develop anywhere along the midline of the body, they are most common in the sacro-coccyx and ovaries [5]. Almost 80% of teratomas occur in women. Morphologically, they can be solid, cystic or mixed tumors. Various degrees of differentiation of teratomas are observed, from very immature neuroectodermal structures to fully differentiated glandular and epithelial tissues, sometimes containing teeth or hair. They are divided into mature teratomas and potentially malignant immature teratomas, most of which in children are mature [6]. In our patient's case, the tumor turned out to be an immature teratoma.

They may contain tumor of the endodermal sinus and, less commonly, chorionic carcinoma. Both types predispose to local recurrences and metastases [7].

These tumors can be monitored by determining the concentration of tumor markers such as AFP and β-hCG, which was ordered in the patient [8].

Craniofacial teratomas are rare neoplasms, accounting for 2% of neonatal tumors, with an estimated frequency of between 1 : 40,000 and 1 : 80,000 live births [5]. They can develop in many craniofacial structures and include the nasopharynx, paranasal sinuses, tongue, tonsils and palate. Sometimes they are also found in the thyroid gland. Often these tumors reach large sizes.

Initial diagnosis of teratomas is based on prenatal ultrasound and is an excellent method of screening. Magnetic resonance imaging is a helpful method in assessing the tumor anatomy and in selecting the method of surgical treatment [9]. In this case it was decided that CT was better suited as preoperative assessment of tumor vascularization.

Early radical surgical resection is the only therapeutic option in case of congenital teratoma and it is performed after the patient's condition is stabilized, preferably within the first 24–48 hours of life [10].

CONCLUSIONS

Thanks to imaging examinations (USG, MR), teratomas can be diagnosed prenatally. This makes it possible to plan a delivery in a center with a high degree of reference, with the possibility of appropriate surgical treatment.

Multidisciplinary cooperation with the participation of neonatologists, anesthesiologists, otorhinolaryngologists and surgeons is very important. Early tumor resection and appropriate perinatal and perioperative care increase the chance of survival of a newborn with craniofacial teratoma.

DISCLOSURE

The authors declare no conflicts of interest.

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