

PRIMARY MATURE MEDIASTINAL TERATOMA IN A NEWBORN – A CASE REPORT

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Summary

Primary mediastinal teratomas, whether mature or immature, are very rare. They could cause serious life threatening respiratory obstruction at newborn age. This report presents clinical course, imaging, autopsy and pathohistological findings in a newborn with mature mediastinal teratoma, which led to severe respiratory failure and death. Despite the fact that postnatal respiratory distress was rarely caused by mediastinal tumor, that type of tumor should be taken into consideration in case of severe perinatal asphyxia. The case is therefore worth of presentation.

KEY WORDS: *teratoma, mediastinum, newborn*

ZRELI PRIMARNI TERATOM MEDIJASTINA U NOVOROĐENČETA: PRIKAZ SLUČAJA

Sažetak

Primarni teratomi medijastina, bez obzira jesu li zreli ili nezreli, rijetkost su i u novorođenačkoj dobi mogu biti uzrokom teške, za život opasne opstrukcije dišnih puteva. U ovom radu prikazan je klinički tijek, pretrage, obdukcijski i patohistološki nalazi u novorođenačeta sa zrelim teratomom medijastina koji je prouzročio težak oblik respiratornog zastoja i smrt. Unatoč činjenici da su postnatalne respiratorne smetnje rijetko prouzročene tumorom medijastina, tu vrstu tumora treba uzeti u obzir u slučaju teške perinatalne asfiksije. Stoga ovaj slučaj vrijedi prikazati.

KLJUČNE RIJEČI: *teratom, medijastin, novorođenče*

INTRODUCTION

Mediastinal tumors at newborn age are very rare and cause compression of vital structures. Teratomas prevail in that region (1, 2). Few cases of primary mature mediastinal teratomas have been described in literature with dominantly severe clinical course, presenting by severe respiratory distress due to compression of vital structures (3). Primary mediastinal germ cell tumors present a particular group of tumors characterized by uncommon histological and clinical heterogeneity (4).

CASE REPORT

We presented a male newborn of 3,880 g birth weight, delivered by a young and healthy mother from the second normal pregnancy. The delivery occurred at 38 weeks gestation due to amniotic rupture, vaginally, with head presentation. No spontaneous breathing occurred at birth. There was bradycardia and heart sounds were heard over the right hemithorax dominantly. Apgar score was 5/5. The baby was admitted to the neonatal intensive care unit following the primary resuscitation. Respiratory de-

pression, superficial and spastic respiration, generalized cyanosis, ongoing bradycardia and heart sounds with punctum maximum heard over the right hemithorax were the principal clinical findings. The newborn was intubated and ventilated mechanically. Umbilical and urinary catheters were inserted. Due to bleeding through the catheters and nasotracheal tube, coagulation tests were done and disseminated intravascular coagulation determined thereupon (prothrombin time 1.68 INR, APTV 53 s, antithrombin III 37.3%, d-dimers 2131 ug/L). Routine laboratory findings showed severe combined metabolic and respiratory acidosis (pH 6.81, pCO₂ 19.6 kPa, pO₂ 8.86 kPa, HCO₃ 22.6 mmol/L, BE -18.9) and marked leukocytosis ($51.3 \times 10^9/L$). The child received combined empirical antimicrobial therapy and infusion of glucose and electrolytes solutes. Metabolic acidosis was corrected, fresh frozen plasma administered, antithrombin III replaced, surfactant and inotropic support applied. An urgent chest x-ray was done following primary stabilization, with neither lungs nor heart appearance, not even after the surfactant application (Figure 1). Cardiac ultrasound excluded an inborn heart defect. Thoracic ultrasound (US) revealed a large tumor mass of mixed solid and cystic appearance in the lower anterior mediastinum occupying the whole left hemithorax, 50x50 mm in diameter, and bilateral pleural effusions. A big cystic mass of almost the same size belonged to the same tumor. As an adequate preoperative clinical stabilization was not achieved,

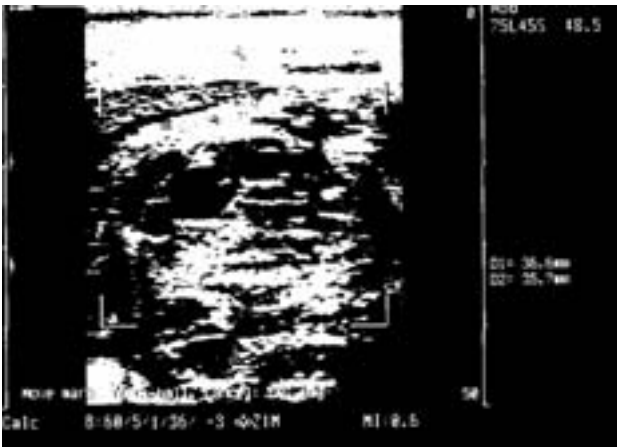


Figure 1. Ultrasound view of the anterior mediastinum with a solid-cystic tumor

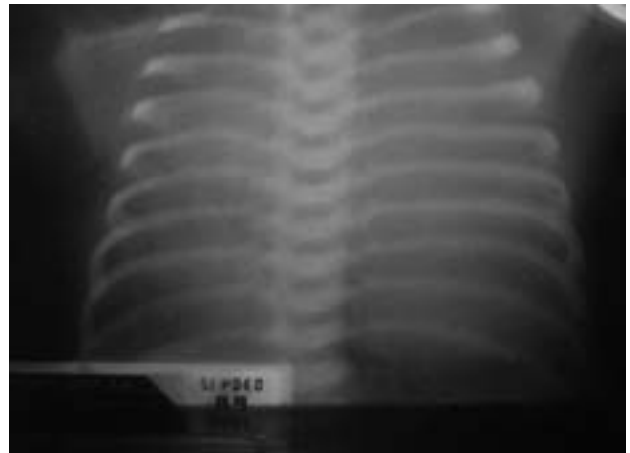


Figure 2. Anteroposterior chest x-ray does not show a functional pulmonary parenchyma

US-guided thoracocentesis was performed in order to render lungs decompression. Fifty milliliters of yellowish fluid containing phagocytes and erythrocytes was drawn from the cyst. Despite the undertaken therapeutic procedures, an optimal oxygenation was not achieved, heart failure ensued gradually, consecutive cardiac arrest and finally lethal outcome occurred after only 18 hours of life.

AUTOPSY REPORT

A male newborn, 54 cm in length and of 3,900 mg weight, was without external anomalies. The thymus was moved rightwards in the upper anterior mediastinum. A tumor smooth in surface, 8.5x7.5x4.5 cm in size, dominated the anterior medial and lower mediastinal space. The left lung was situated behind the tumor in the left hemithorax, while the heart with the pericardium was moved rightwards compressing the right lung. The described tumor did not belong to any thoracic structure, i.e. all thoracic structures were slightly separated from the mass. The tumor was both solid and cystic, and cystic holes were filled with serous yellowish, somewhat mucous fluid. Both lungs were reduced in volume and in air content, with smooth pleura, which was imbued by dotted and patchy bleedings. Both lungs were anatomically and morphologically without anomalies. An important finding was an extremely narrowed trachea at bifurcation due to external compression by the tumor (Figure 3).

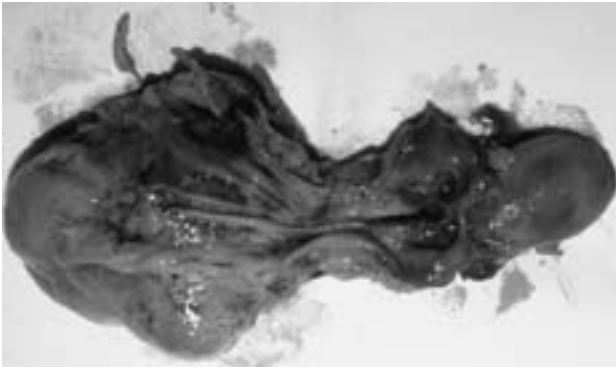


Figure 3. Macroscopic appearance of a solid and cystic mediastinal tumor

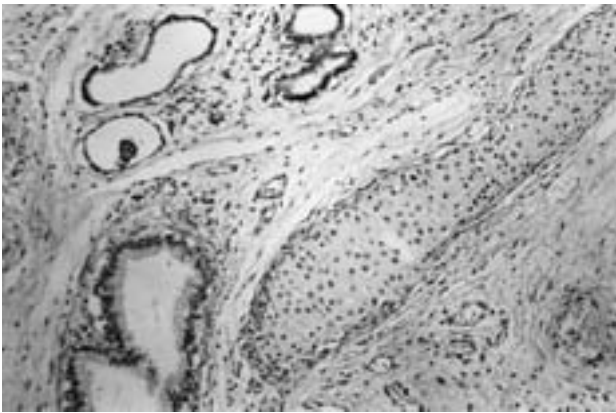


Figure 4. Microscopic view of the tongue and mediastinal structures with the open trachea showing an obstruction in the bifurcation portion

Cardiac section revealed dilated right heart cavities and right ventricular wall hypertrophy, which was 3 to 7 mm thick. The remaining autopsy findings were normal. Histologically, the tumor consisted of the three germ leaves elements. Cystic spaces predominated, lined by cylindrical mucous epithelium, respiratory and multilayer planocellular epithelium. Solid regions of hyaline cartilaginous insula were found between the cysts, as well as the brain tissue with parts of the chorioid plexus, and the exocrine gland tissue - salivary and pancreatic. Structures histologically corresponding to the eye stem tissue were found, too. All tumor components were mature (Figure 4). Other organs showed generalized congestion, and particularly significant was intraalveolar and subpleural bleeding findings, as well as foci of extramedullar hematopoiesis in the liver.

DISCUSSION

Teratomas are the most frequent neonatal neoplasms, usually benign and mostly located in the sacrococcygeal region (5). Mediastinal teratomas are uncommon in newborns and in children, corresponding to 7-10% of all teratomas in that age group (6). Even though large tumors can be asymptomatic, teratomas are well known for respiratory distress at neonatal age due to vital structures compression and consecutive acute and chronic respiratory system pathology (1, 7). The only therapy is surgery, although acute compression requires urgent thoracotomy. Surgically treated teratomas at that age were usually benign. The treatment prognosis at that age depends on the grade of success of airways patency restoration (7).

A suspicion of a mediastinal teratoma comes from a mediastinal mass by radiography, which nature is to be determined by US or computed tomography. In our case the chest x-ray was negative, i.e. the mass was not shown up. However, functional pulmonary parenchyma was not found by that method either, which indirectly indicated compression. Differential diagnosis consists of hamartomas, thymomas, germinomas, mediastinal lymphomas and vascular aberrations (8). The literature review showed respiratory distress as the most common presentation of mediastinal teratomas in newborns (3). As in our patient, functional pulmonary parenchyma was not found by chest x-ray imaging, the child presented with respiratory distress followed by bleeding, disseminated intravascular coagulation according to coagulation tests abnormalities with predominantly high D-dimers value, and without septic signs by physical examination. The assessment of D-dimer concentration is an essential part in the diagnostic procedure of thromboembolic diseases. D-dimers are the products of fibrin hydrolysis with elevated levels in fibrinolytic processes. In clinical practice, problems exist in the interpretation of increased D-dimer levels, especially without thromboembolic disease. Before starting further expensive imaging diagnostics, other reasons such as pregnancy, neoplasma, systemic inflammatory disease, should be considered in differential diagnosis (9). Ultrasound revealed the expansive mediastinal process.

Histological classification of teratomas dividing them into mature, immature and malignant has been well accepted. Mature teratomas have differentiated elements and benign clinical nature with the exception of mediastinal tumors. Germ cell tumors in males stem from testicles, but a small proportion, 2-5% is of the extragonadal origin. Extragonadal germ cell tumors are histologically identical to their gonadal peers, but, unlike gonadal tumors, are situated along the median line of body. Extragonadal germ cell tumors are considered to be a consequence of malmigration of germ cells along the urogenital fissure during embryogenesis, or a result of physiologic distribution of germ cells into liver, bone marrow and brain, in order to ascertain normal function and transmission of hematological and immunological information (10).

CONCLUSION

A case of a large congenital mature mediastinal teratoma was presented. The tumor caused insufficient tracheal bifurcation and bronchial tree development, and led to the compression of both lungs and heart movement into the right hemithorax followed by right ventricular hypertrophy (cor pulmonale). Until 1980, there were only 5 cases of mediastinal tumors in newborns reported in the literature. During the past 20 years, the largest study presented 15 patients, 6 of them newborns with mediastinal teratomas. The mediastinum is undoubtedly a rare site of teratoma in newborns, and if this is the case, immature forms prevail, mostly clinically presenting by respiratory distress resulting from the vital structure compression (3). Our patient experienced the compression of vital structures as early as *in utero*, followed by pulmonary hypoplasia, rendering the undertaken therapeutic procedures unsuccessful in establishing adequate oxygenation, leaving the patient unstable for surgical treatment. Based on this experience, the suggestion to take this possibility into consideration

in the differential diagnosis of respiratory distress seems reasonable.

REFERENCES

1. Allman AW, Buss PW. Mediastinal teratoma presenting as apparent fresh stillbirth. Arch Dis Child Fetal Neonatal ED 2001;84:F65-F66
2. Das BB, Richardson C, Lala R, Sahoo. Primary immature mediastinal teratoma in a newborn. Indian Pediatrics 2001;38:1181-4.
3. Lakhoo K, Boyle M, Drake DP. Mediastinal teratomas: Review of 15 pediatric cases. J Pediatr Surg 1993;28:1161-4.
4. Schneider DT, Calaminus G, Reinhard H, Gutjahr P, Kremens B, Harms D, Göbel U. Primary mediastinal germ cell tumors in children and adolescents: Results of the German cooperative protocols MAKEI 83/86, 89, and 96. J Clin Oncol 2000; 18:832-9.
5. Stivens MCG. Neonatal tumours. Arch Dis Child 1988;1122-5.
6. Tapper D, Lack EE. Teratomas in infancy and childhood. A 54-years experience at the Children's Hospital Medical Center. Ann Surg 1983;198:398-410.
7. Mogilner JG, Foncesa J, Davies MRQ. Life-threatening respiratory distress caused by a mediastinal teratoma in a newborn. J Pediatr Surg 1992;1519-20.
8. Whittaker LD, Lynn HB. Mediastinal tumors and cyst in the pediatric patient. Surg Clin North Am 1973;53:893-904.
9. Ziegler T, Murzik M, Schau A, Winkler C, Funfstuck R. Interpretation of increased D-dimer values. Hamostaseologie 2004;24(2):144-6.
10. Bokemeyer C, Nichols CR, Droz JP, Schmoll HJ, Horwich A, Gerl A, Fossa SD, Beyer J, Pont J, Kanz L, Einhorn L, Hartmann JT. Extragonadal germ cell tumors of the mediastinum and retroperitoneum: Results from an international analysis. J Clin Oncol 2000; 20:1864-73.

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