Congenital mediastinal immature teratoma: a case report with autopsy findings

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SUMMARY: Aksoy F, Şen C, Danışment N. Congenital mediastinal immature teratoma: a case report with autopsy findings. Turk J Pediatr 2002; 44: 76-79.

A full-term newborn with karyotype 46, XX was delivered by cesarean section. She had severe respiratory distress and substernal retraction, and underwent emergency operation, but she died on the same day due to respiratory failure.

The mother, 26-year-old prima gravida with no history of twinning, had been examined with ultrasonography at the 34th week of her pregnancy, which revealed a fetus with edema of head and neck region, a probable diaphragmatic hernia, polyhydramnios, and a large mediastinal mass with solid and multicystic parts with hypoplasia of the lungs.

Autopsy revealed a 9 x 5 x 3 cm lobulated mediastinal mass with both solid and cystic areas, displacing the lungs and the heart postero-inferiorly and thymus anteriorly. The lungs were hypoplasic. Microscopically, the mass showed mature epithelial and mesenchymal tissues with primitive mesenchyme and immature neuroepithelium. All these findings led to the diagnosis of an immature teratoma.

Mediastinal teratomas are rare and life-threatening, but early diagnosis and surgical intervention in a newborn with sufficient lung maturation may provide a long survival.

Key words: mediastinal teratoma, extragonadal teratoma, immature teratoma.

Mediastinal teratomas account for up to 20% of mediastinal masses in children. However, presentation in the neonate is rare, with only five cases reported in the literature before 1980[1-3].

The anterior mediastinum, the pericardium and, rarely, the lungs are principal sites of teratomas in the thoracic cavity. Respiratory distress and chest pain are the common presenting symptoms. Unusual manifestations include hemoptysis and hypoglycemia. Some affected children have no symptoms[4-8].

Mature and immature teratomas are the basic pathologic types of teratomas which mostly occur in the mediastinum and pericardium[3-4].

Here we present the clinical features and autopsy findings of a newborn with congenital mediastinal immature teratoma which caused a fatal respiratory distress.

Case Report

A 26-year-old prima gravida with no family history of twinning underwent cordocentesis at 34 weeks' gestation, and fetal blood sampling revealed karyotype of 46, XX. During this procedure polyhydramnios was noted and 1000 ml of amniotic fluid was evacuated.

A term baby girl with an APGAR score of 4 at 5th minute was delivered by cesaren section. There was severe respiratory distress and substernal retraction at the time of birth. She was duly transferred to the Pediatric Surgery Ward of the hospital for an emergency operation, but died on the same day due to respiratory failure.

Ultrasonographic Findings: An ultrasonographic examination at 34 weeks' gestation showed edema in the head and neck region, a probable diaphragmatic hernia, polyhydramnios, and a large solid multilocular cystic mass in the mediastinum with hypoplasia of the lungs.

Autopsy Findings: A full-term baby girl with normal growth and development was seen. No gross abnormality was observed, except for focal edema in the head and neck.

When the thorax and abdomen were dissected, a huge, lobulated, solid, partially cystic mass was seen, filling the mediastinum and displacing

the lungs and heart postero-inferiorly and the thymus anteriorly. The lungs were hypolasic. The rest of the organs were found to be hyperemic and showed no special features.

Gross Appearance : A 9 x 5 x 3 cm large mass was found with a multilobular outer surface, covered by a thin capsule. Cut surface showed multiple cysts with shiny solid and soft mucoid areas. The cysts varied in size from a few milimeters to several centimeters in diameter and were filled either with clear-mucinous or with brown-red fluid (Fig. 1).

Microscopic Findings : Small cystic and solid areas were quite distinct in the tumor. Cysts, in general, were lined by either single layered or stratified, columnar and mucin-secreting epithelia. The walls of some of the cysts resembled intestinal and bronchial mucosa (Fig. 2). In between and around the cysts were mature mesenchymal tissue, glandular elements, islands of cartilage, primitive brain tissue and scattered immature neuroepithelium (Figs. 3, 4). The tumor, which showed features of immature teratoma, contained mature tissue, immature mesenchyme and neuroepithelia tissues from all three germ layers; hence, our case was diagnosed as immature teratoma.

Discussion

Mediastinal teratomas are rare neoplasms in children, accounting for only 7% of all germ cell tumors. The presentation of mediastinal teratoma in the newborn is sparsely documented. In three series there were only four neonates in a total of 28 children. In the Lakloo[1] series, 40% (6/15) of the patients were neonates. In our series of 47 teratomas, 19 were in neonates and only one had mediastinal localization[9].

Teratomas are complex tumors composed of tissues originating from all three germinal layers[4,8,10-13].

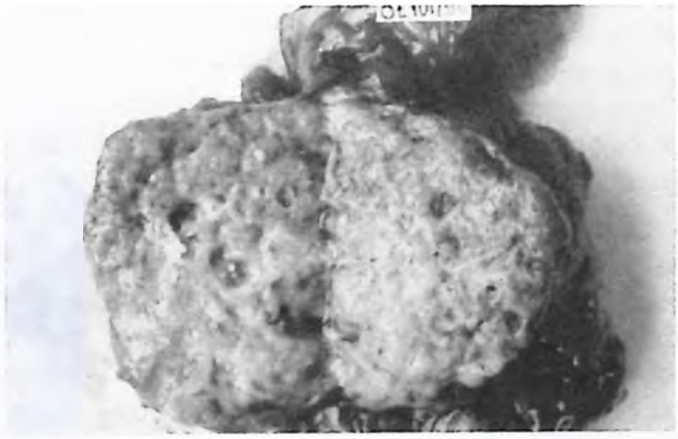
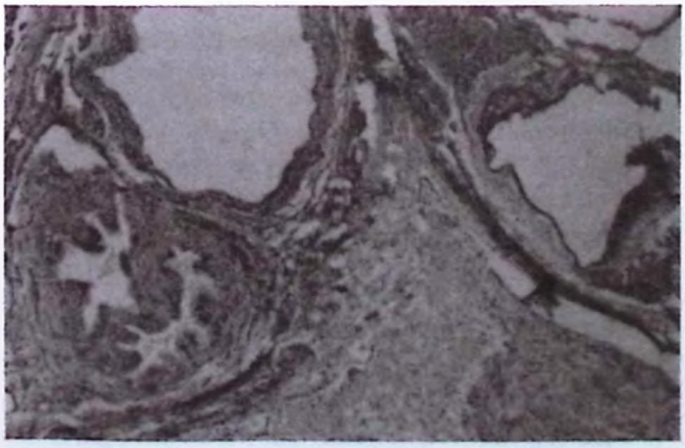


Fig. 1. The multilobulated with small cystic areas containing fluid and shiny solid fibrous areas with soft mucoid regions.



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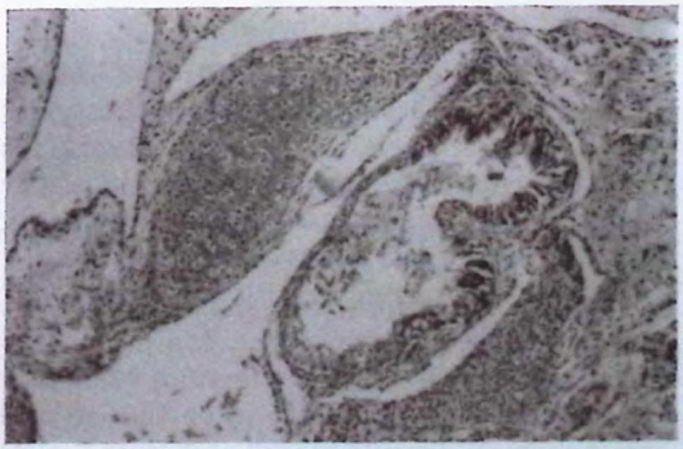

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Fig. 2, 3. The histological appearance: The tumor contained various epithelial components such as cubic, stratified squamous epithelium, and bronchial mucosa type epithelium lining the cysts. Surrounding these cysts were mature cartilage, immature neuroepithelium and primitive mesenchyme.

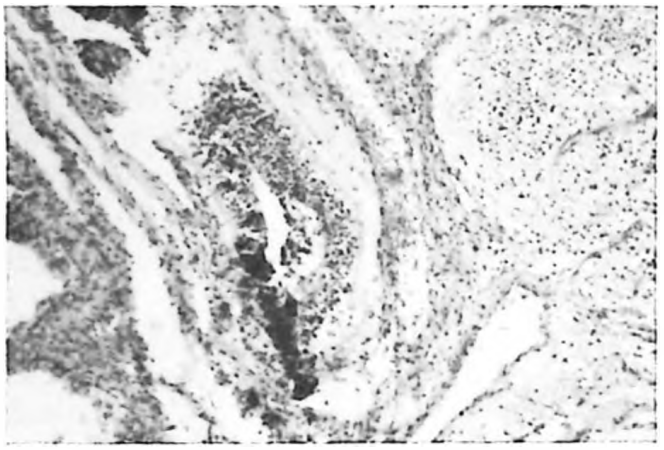

Fig. 4. Primitive mesenchyme and embryonic neuroepithelial tissue (primitive neural tissue component) were noted in other areas.

They usually occur in a para-axial or mild-line location from the brain to the sacral area and in gonads[8,12]. Less common sites include the posterior mediastinum, pericardium, heart, and lung parenchyme[1].

Mediastinal teratoma in a newborn may cause severe respiratory distress which is the most important clinical finding and requires immediate intubation and surgical intervention[6-8,12,14]. Respiratory distress was the main clinical presentation in our case but the time was insufficient for surgical intervention.

The congenital teratomas, if diagnosed and treated shortly after, birth, are usually benign regardless of the somatic immaturity; on the other hand, if the tumor has a critical site such as the central nervous system, the pericardium or mediastinum, it may cause death by local effects[4].

The prenatal diagnosis of a mediastinal teratoma ultrasonographically is based on a tumor composed of cystic and solid structures in the region of the upper mediastinum[8].

Differential diagnosis should include all intrathoracic malformations such as bronchogenic cysts, congenital adenomatoid malformation of the lungs, hamartoma of the lungs, bronchopulmonary sequestration, diaphragmatic hernias, thymoma, lymphoma and cystic hygroma[1,7,8].

Mediastinal teratomas have been classified as mature when there is histologically well differentiated tissue and as immature when they contain so-called immature epithelial and mesenchymal elements as well as mature tissues (especially tissues of neuroepithelia). The malignant group of immature teratomas consists of embryonal carcinomas, yolk sac tumor, seminoma and choriocarcinoma[1,2,10].

In the present case, we observed foci of mature elements containing primitive mesenchyme and primitive neuroepithelial tissues scattered through the mass (Fig. 4).

In such cases α-fetoprotein has been indicated as a good marker, which is especially valuable in the follow-up of patients for tumor recurrence[1].

The origin of the extragonadal teratoma is considered to be different from that of the gonadal. Extragonadal teratomas arise from early embryonic cells of primordial germ cells in the course of migration during embryogenesis. One hypothesis suggests that extragonadal lesions are often congenital and are typically misplaced in the midline in conjoined twin pregnancies, whereas gonadal teratomas may arise from sequestered haploid germ cells[10].

Since the teratomas show rapid growth in early gestational periods, they are responsible for the compression of the neighboring organs. Mediastinal teratoma, depending on the size, might cause polyhydraminos or fetal hydrops, because of a depressed esophagus or the obstruction of the venous return[15].

Diffuse edema in the head and neck region and polyhydramnios may indicate a mediastinal mass and, although rare, teratomas in this location should be included in the differential diagnosis when these findings are present in ultrasonography.

These masses are life-threatening, but early diagnosis and immediate surgical intervention may provide long survival in a newborn with sufficient lung maturation.

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