HK J Paediatr (new series) 2011;**16**:125-127

A Large Mediastinal Immature Teratoma Presenting with Life Threatening Respiratory Distress in a Newborn

M *Mutlu*, N *Kurucu*, Y *Aslan*, B *Saygin*, H *Sarihan*, Ş *Ersöz*

Abstract A 42-hour-old male newborn was referred to our hospital with the complaints of life-threatening respiratory distress. A large mass was observed on posterior-anterior chest radiography. A 10 x 10 cm mass was completely excised from the anterior mediastinum and right hemitorax via right thoracotomy. Histopathologic examination of the mass revealed as immature teratoma. Mediastinal immature teratoma may cause life threatening respiratory obstruction and must be promptly diagnosed and treated.

Key words Immature teratoma; Mediastinum; Newborn; Respiratory distress

Introduction

Primary immature mediastinal teratomas are very rare in neonatal period and constitute 1% of all mediastinal teratomas.[1] The most common presenting symptom of immature teratoma of mediastinum is respiratory distress in newborns.[2,3] If these tumors are promptly diagnosed and treated, their prognosis is excellent. A case of neonate with an immature mediastinal teratoma presenting with life threatening respiratory distress is herein presented.

Faculty of Medicine, Department of Pediatrics, Karadeniz Technical University, Trabzon, Turkey

M Mutlu *MD*
N Kurucu *MD*
Y Aslan *MD*
B Saygin *MD*

Faculty of Medicine, Department of Pediatric Surgery, Karadeniz Technical University, Trabzon, Turkey

H Sarihan *MD*

Faculty of Medicine, Department of Pathology, Karadeniz Technical University, Trabzon, Turkey

Ş Ersöz *MD*

Correspondence to: Dr M Mutlu

Received October 12, 2010

Case Report

A 42-hour-old boy was referred to our neonatal intensive care unit with complaint of respiratory distress. He had been born by cesarean section at 34 weeks' gestation, 2700 gram birth weight, to a 36-year-old mother (G5,P2). Resuscitation had been applied at delivery and respiratory support had been given with nasal continuous positive airway pressure to the neonate. Physical examination revealed severe respiratory distress, marked by grunting, intercostal and subcostal retractions, and cyanosis. There was no haemodynamic instability. The arterial blood gas showed pH: 6.94, pCO_2: 69 mmHg, pO_2: 49.6 mmHg, HCO_3: 14.7 mmol/L, BE:-17.4 mmol/L, and SpO_2: 73.8. The patient was intubated and respiratory support was given with mechanical ventilation in prone position. Cyanosis and blood gas improved followed intubation and ventilation. A large mass was observed on posterior-anterior chest radiography (Figure 1). Serum α fetoprotein (AFP) level was 17,864 U/ml (425-55,270 U/ml). Beta-human chorionic gonadotropin (β-HCG) and vanillylmandellic acid were normal. Computed tomography (CT) scan demonstrated a solid mass with heterogen density and no calcification that completely filled to anterior mediastinum extended through the right hemithorax, compressed to the main vascular structures and trachea, and pushed the heart to the left side (Figure 2). A 10 x 10 cm solid mass was completely excised from the anterior mediastinum and right hemithorax through

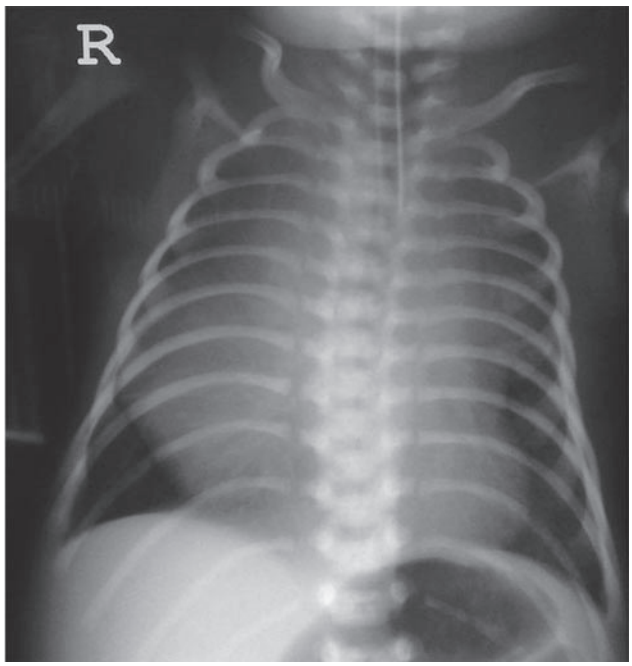

Figure 1 Posterior-anterior chest radiography shows a mediastinal enlargement.

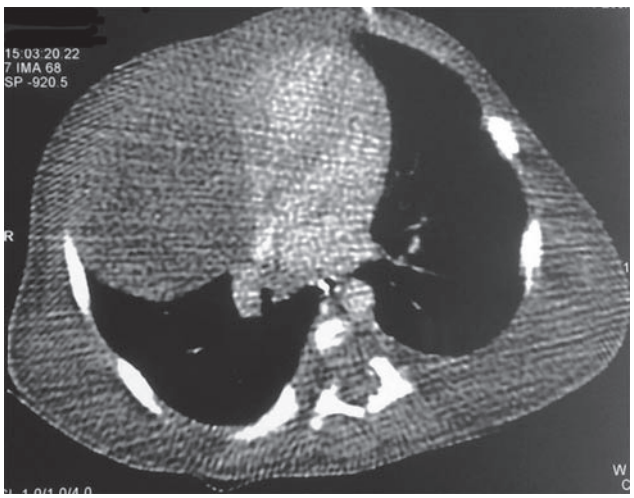

Figure 2 Chest CT shows an anterior mediastinal mass extending into right hemithorax.

a right anterolateral thoracotomy. Histopathologic examination of the mass revealed mature and immature neuroepithelial elements, glandular columnar epithelium, cartilage, rosette formation, and cystic structures in the mesodermal connective tissue and diagnosed as immature teratoma, consistent with a grade II (Figure 3). We accepted our case as stage 1 as the tumor was completely resected and there were no regional lymph node involvement or

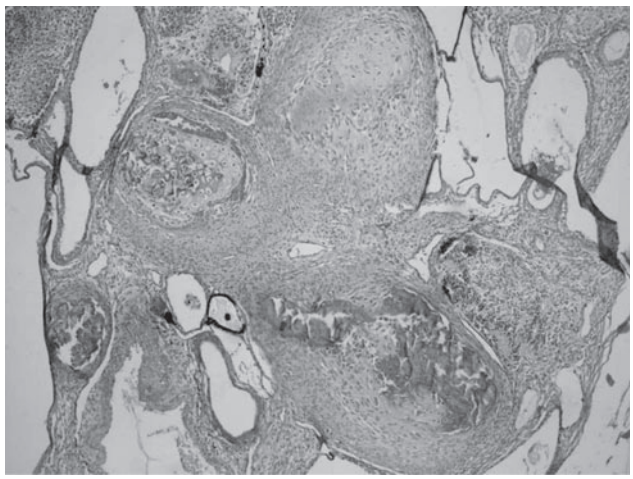

Figure 3 Neoplasm consist of predominantly mesodermal elements accompanied by multiple minute cysts and neuroepithelial tubular structures (HE x100).

distant metastasis. The AFP levels remained within normal limits at regular intervals in postoperative follow-up and no recurrence has been observed for two years.

Discussion

Teratomas are embryonic neoplasm arising from primordial germ cells. They are classified as either mature or immature. Congenital teratomas are commonly immature. Teratomas may cause high percent of morbidity and mortality. Most of the congenital teratoma are extragonadal and are located frequently in the sacrococcygeal region in neonate. In our case tumor was located in mediastinum. Mediastinal teratoma accounts for approximately 2.4-10% of all teratomas in children, and less than 1% of neonatal period.[1-3] Immature teratomas are more common in mediastinum than mature ones.[3]

Mediastinal teratomas usually occur within the anterior mediastinum. Mediastinal teratomas in infancy usually cause compression of vital structures. Congenital mediastinal teratomas may be presented with respiratory distress, hydrops fetalis, polyhydamnios and stillbirth.[1-4] Respiratory distress is the most common presentation of immature teratoma of mediastinum in newborn.[2] Respiratory distress usually occurs due to compressing of the airway and/or hypoplasia of the lung. Our patient presented with life threatening respiratory distress due to tracheal compression. Aggressive respiratory and cardiovascular management may be required.[4] Aggressive

respiratory management is required in our patient. We ventilated our patient in prone position as to decrease the compression of tumor to the vital structures. Compression of tumour to the vital structures may be decreased in prone position in patients with tumor localised in the anterior mediastinum.

Immature teratomas occurring in the fetus and newborn are associated with a more favourable prognosis than in adolescents and adults.[5,6] Total surgical removal is curative. McKenney et al[5] reported that the clinical behavior of congenital teratomas is determined predominantly by whether or not the tumor can be completely resected and did not correlate with grade and presences endodermal glands. In our case complete surgical excision of the tumor could be performed. Perinatal teratomas have a relatively low (5%) recurrence rate.[3] AFP has an important role in initial diagnostic evaluation, and follow-up of recurrence.[5] Serum AFP level is high at birth and decreases to normal adult levels within the first 8-10 months.[7] This situation should be remembered for the diagnosis and follow-up. In our patient, AFP levels were normal limit in initial diagnosis and follow up. Recurrence has not been observed in our patient until now.

Imaging studies are important for diagnosis. Mediastinal teratoma usually manifests on CT as a heterogeneous anterior mediastinal mass containing soft-tissue, fluid, fat, or calcium attenuation, or any combination of the four.[8] In our case, CT scan showed a heterogeneous anterior mediastinal mass containing soft-tissue with no calcification.

The diagnosis of immaturity is most commonly based on the presence of immature neuroepithelial structures and the other immature elements including cartilage and glandular epithelium.[4] In our case mature and immature neuroepithelial structures, cartilage, glandular epithelium were observed on histopathologic examination.

Differential diagnoses with other anterior mediastinal masses such as tymoma, thymic cyst, lymphoma, lymphangioma, lipoma, bronchial and enteric cyst, and neurogenic tumours.[9]

We suggest that, although mediastinal immature teratoma is seen rarely in neonatal period, it may cause life threatening respiratory obstruction and aggressive respiratory and cardiovascular management may be required. Respiratory distress may be decreased by applying ventilation in prone position. It must be promptly diagnosed and treated.

References

1. Das BB, Richardson C, Lala R, Sahoo S. Primary immature mediastinal teratoma in a newborn. Indian Pediatr 2001;38: 1181-4.
2. Lakhoo K, Boyle M, Drake DP. Mediastinal teratomas: review of 15 pediatric cases. J Pediatr Surg 1993;28:1161-4.
3. Isaacs H Jr. Perinatal (fetal and neonatal) germ cell tumors. J Pediatr Surg 2004;39:1003-13.
4. Allman AW, Buss PW, Spicer RD, Wake A. Mediastinal teratoma presenting as apparent fresh stillbirth. Arch Dis Child Fetal Neonatal Ed 2001;84:F65-6.
5. McKenney JK, Heerema-McKenney A, Rouse RV. Extragonadal germ cell tumors: a review with emphasis on pathologic features, clinical prognostic variables, and differential diagnostic considerations. Adv Anat Pathol 2007;14:69-92.
6. Carter D, Bibro MC, Touloukian RJ. Benign clinical behavior of immature mediastinal teratoma in infancy and childhood: report of two cases and review of the literature. Cancer 1982;49:398-402.
7. Tsuchida Y, Endo Y, Saito S, Kaneko M, Shiraki K, Ohmi K. Evaluation of alpha-fetoprotein in early infancy. J Pediatr Surg 1978;13:155-6.
8. Moeller KH, Rosado-de-Christenson ML, Templeton PA. Mediastinal mature teratoma: imaging features. AJR Am J Roentgenol 1997;169:985-90.
9. Taneja L, Chadha R, Singh D, Chouudhury SR. Massive anterior mediastinal teratoma in an infant. J Indian Assoc Pediatr Surg 2004;9:206-8.