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Mediastinal Teratoma In A Newborn Presenting With Respiratory Distress And Superior Mediastinal Syndrome: A Case Report

Yenidoğanda Solunum Güçlüğü Ve Süperior Mediasten Sendromuna Sebep Olan MediastinaL Teratom: Olgu Sunumu

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Özet

Amaç: Mediastinal teratomlar, çocukluk çağı mediastinal tümörlerinin %20'sini, tüm teratomların ise %10'unu oluştururlar ve yenidoğan döneminde solunum güçlüğünün nadir nedenlerindendir.

Metod: Solunum güçlüğü ve superior mediasten sendromuna sebep olan bir yenidoğan mediastinal teratom olgusu sunduk. Sağ akciğer üst loba komşu 6x4x2,5 cm'lik kitle total olarak çıkartıldı.

Bulgular: Histopatolojik değerlendirmede sağlam bir kapsülle çevrili, immature kıkırdak, mezenkim ve nöroepitel dokuları içeren, ever 2 immatür teratom izlendi. Kemoterapi verilmedi. 6. Ay kontrolünde, AFP düzeyi 18 ng/ml ölçüldü ve yaşıyla uyumluydu. Rekürans görülmedi.

Sonuç: İyi huylu mediastinal teratomların çoğu yavaş büyürler ve çoğu erken puberteye kadar semptom vermezler. Direk grafi, toraks ultrasonografisi, bilgisayarlı tomografi ve manyetik rezonans, kitlenin yerinin ve çevre dokularla ilişkisinin değerlendirilmesinde yardımcıdır. 15 yaşın altındaki çocuklarda mediastinal teratomların tedavisi total cerrahi eksizyondur.

Anahtar Kelimeler: Yenidoğan, solunum güçlüğü, superior mediasten sendromu, teratom.

Abstract

Background: Medistinal teratomas compose 20% of childhood mediastinal tumors, 10% of all teratomas and it is an uncommon cause of respiratory distress in neonatal period.

Method: We report a newborn with mediastinal teratoma presenting with respiratory distress and superior mediastinal syndrome. The 6x 4x 2,5 cm mass adjacent to the right upper lung was totally resected.

Results: Histopathologically Grade 2 immature teratoma with an intact looking capsule, and components of immature cartilage, mesenchymal and neuroepithelial tissue was identified Chemotherapy was not administered. At the 6^{th} month of her follow up, the AFP level was 18 ng/ml in concordance with her age. No recurrence was observed.

Conclusions: Most of benign mediastinal teratomas grow slowly and do not manifest symptoms until early puberty. Chest X-ray, thorax ultrasonography, computed tomography and magnetic resonance imaging are useful in identifying the localization and association of mass with peripheral tissues. Treatment of the mediastinal teratomas in children under the age of fifteen is total surgical excision.

Key Words: Newborn, respiratory distress, superior mediastinal syndrome, teratoma.

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INTRODUCTION

Mediastinal teratoma is an uncommon cause of respiratory distress in neonatal period. [1] It composes 20% of childhood mediastinal tumors and 10% of all teratomas. [2,3] Primary immature mediastinal teratomas, composing approximately 1% of mediastinal teratomas are very rare. [4]

In current study, we report a newborn with mediastinal teratoma presenting with respiratory distress and superior mediastinal syndrome.

CASE REPORT

A 16 day old female neonate, born at 35 week of gestation as a twin sibling by cesarean section and weighed 2150 g. It was admitted to a hospital at 14 days of age and considered to have aspiration pneumonia because of respiratory distress. Antibiotic treatment was started. But her respiratory distress progressed and patient was referred to our intensive care unit. Her twin sibling did not have any medical problem. On physical examination, she had a respiratory rate of 70/min, a body temperature of 36,3 °C and a pulse rate of 144/min, and her blood pressure was 90/60 mmHg. She was dyspneic, tachypneic and she had subcostal, intercostal retractions. She had edema over her face and neck (Figures 1). Electrolytes and complete blood count results were normal. C reactive protein was negative. In arterial blood gas analysis was as follows; pH:7,31, pCO₂: 58 mmHg, pO₂: 96 mmHg, O₂ saturation 97%, HCO₃:19 mmol/L, base excess 2 mmol/L. The patient was intubated and required mechanical ventilation. Upon seeing an uniformly ounded consolidation suggesting a mass on her postero-anterior chest X-ray (Figure 2), an ultrasonography (USG) was performed. On the

apical region of the right lung, a 31x40 mm sized hypoechoic solid lesion extending to left with cystic area, was observed suggesting cystic adenomatoid malformation or teratoma.

On chest computed tomography (CT) images (Figure 3), a 30x30x38 mm sized uniformly bounded mass containing cystic and solid components, adjacent to the posterior thymus, displacing the vasculature to the periphery and compressing the trachea was seen in the right side of the superior mediastinum.



Figure 1: The edematous face and neck of baby (superior mediastinal syndrome)

Human chorionic gonadotropin level (β HCG) was <2 ng/ml (N: < 2 ng/ml) and alpha fetoprotein (AFP) level was 20100 ng/ml (N: 9452 ± 12610 ng/ml). On echocardiographic examination, the cardiac functions were all in normal limits, whereas turbulance was detected in the pulmonary and systemic veins due to compression of

the tumor. The 6x 4x 2,5 cm mass adjacent to the right upper lung was totally resected (Figure 4). Histopathologically Grade 2 immature teratoma with an intact looking capsule, and components of immature cartilage, mesenchymal and neuroepithelial tissue was identified (Figure 5).



Figure 2: The right upper mediastinal mass in chest X-ray

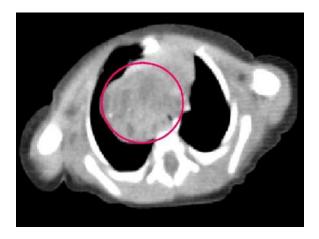


Figure 3: Mediastinal heterogenous mass in thorax tomography (red circle)

The edema over the face and neck was decreased after surgery. She was seperated from the ventilator on day 7th day of admission and was discharged on postoperative day 10. Chemotherapy was

not administered. At the 6^{th} month of her follow up, the AFP level was 18 ng/ml (N: 12.5 ± 9.8 ng/ml) in concordance with her age. No recurrence was observed.



Figure 4: Macroscopic view of teratoma.

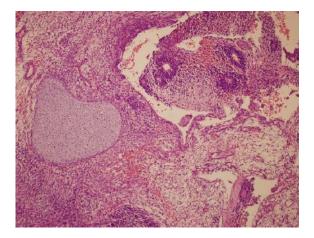


Figure 5: An immature teratoma containing immature chondroid, mesenchyme and neuroepithelial tissue (H&E, x100).

DISCUSSION

Many benign mediastinal teratomas grow slowly and they do not manifest symptoms until puberty, therefore they are often diagnosed by routine chest radiographs. [3] Pain, dyspnea, and cough are the most common symptoms. Our patient had dyspnea and edema over the face and neck. So all of these symtoms gave us the impression of the upper mediastinal mass.

Chest X-ray, thorax USG, CT and MRI are useful diagnostic tools in identifying the tumor's localization and association with peripheral tissues. In current case, a uniformly bounded opacity was seen in the chest radiograph and diagnosis of teratoma was made by further radiological investigations.

The prenatal diagnosis of the mediastinal teratomas are possible. Intrapericardial teratoma. rabdomyosarcoma, cystic adenomatoid malformation. diaphragmatic hernia. pulmonary sequestration, and bronchogenic cyst should also be considered in the differential diagnosis. Our patient had no problems in the antenatal period, and was presented with superior mediastinal symptoms on day 16 after birth.

Serum AFP and β HCG levels are essential for the evaluation and follow up. In our case, the AFP levels were measured during the follow-up and were normal for her age.

Histopathologically teratomas are made of 3 germ layers, ranging from primitive somatic tissues (immature) to organized forms (mature). Immature teratomas grow more quickly than the mature ones and they predominantly contain neuroepithelial elements. However malignancy is rarely seen in childhood. In the pathological examination of our patient, an encapsulated tumoral constitution made of immature mesenchymal and glandular elements, containing multiple cystic and solid components was seen. The treatment of choise for the mediastinal teratomas in children under the age of fifteen is total surgical excision. [5] There is no indication for chemotherapy after the total resection of the tumor as seen in our patient. For the with incomplete patients resection containing volk salk components microscopically, relapse is likely at any age. [6] During 6 months of follow up, no recurrence was observed in our patient. For the newborns presenting with respiratory distress and superior mediastinal syndrome, mediastinal tumors should be considered and essential imaging techniques need to be performed. [7]

In conclusion, mediastinal teratoma presenting with respiratory distress and superior mediastinal syndrome in newborn are very rare and, they should be considered in newborns presenting with respiratory distress. Such patients should be followed up using multidisciplinary modality in terms of long term recurrence.

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