Case report

Neonatal neuroblastoma presented with respiratory distress, a case report

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A R T I C L E  I N F O

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A B S T R A C T

Background: The most common neonatal tumor is neuroblastoma and adrenal gland is the most common site of involvement. 11–26% of this tumor is seen in the thorax of neonates. Due to a similar appearance of the mediastinal tumor with pneumonia, a high index of suspicion is necessary for early diagnosis of this disorder.

Case report: A 17-day-old female and term neonate delivered by caesarian section was reported in the study. The mother had a normal pregnancy without any reported complications. The newborn admitted to a neonatal intensive care unit (NICU) in her hometown, because of respiratory distress, stridor, and tachypnea which was started from 7 days of life and due to lack of recovery and persistent respiratory symptoms, the patient referred to our hospital. In chest X-ray, opacity in the right upper lobe of the lung was seen. In barium study, a mass like lesion in the size of 35.34 mm adjacent to upper intra-thoracic esophagus with mild mass effect was observed and in CTS a posterior mediastinal mass with severe compression on the trachea and extension to the spinal canal was reported. Due to severe obstruction on the airway, the baby underwent emergency surgery and a mediastinal mass with adhesion and involvement of the ribs and spinal cord was resected. The final diagnosis of poorly differentiated neuroblastoma was confirmed by biopsy.

Conclusion: In every neonate with persistent respiratory distress, stridor and abnormal chest X-ray, diagnosis of neonatal neuroblastoma should be considered.

1. Introduction

Neuroblastoma is an embryonic nervous system tumor in fetal and neonatal periods. Primordial neural crest, which subsequently develops into sympathetic ganglia and the adrenal medulla, is the origin of this tumor. As a result, neuroblastoma can extend along the sympathetic chain anywhere from the neck to the groin [1]. The most common neonatal tumor is neuroblastoma with an incidence of 0.61 per 100,000 live births and accounts for 28–39% of all malignancy in the first month of life [2]. The most common site of presentation in neonatal neuroblastoma is adrenal gland (in 90%) [3]. Another less common form of this disorder is thoracic involvement in 11–26% (4). The prognosis of this embryonic tumor in neonates is good and in most cases, favorable biological characteristics are seen, even despite the metastatic invasion, spontaneously regression is expected [5].

Regardless of a low prevalence of malignancy and neuroblastoma in neonates [6], in unusual neonatal manifestations or abnormal x-ray, the high index of suspicions to this disorder as the most common type of malignant tumor in this period of life is essential.

Neonatal pneumonia is a common underlying disorder of respiratory distress in the first days of life [7]. Based on the postnatal age of the neonates, pneumonia divided into early and late-onset form, [8]. Microbial pattern responsible for both kinds of disease consisted of bacterial, viral, fungal and some protozoa’s that transmitted to neonates from the mother of the baby in early-onset form and from the community in the late-onset form of the disease [9].

Although in some complicated cases of neonatal pneumonia, it needs other diagnostic modalities than chest X-ray, in most of the patients’ clinical symptoms of respiratory compromise besides radiological finding confirm the diagnosis. Differential diagnosis of the persistent right upper and middle lobe pneumonia in a newborn, consisted of tracheoesophageal fistula (TEF), gastroesophageal reflux disease (GERD), pulmonary sequestration, lung hypoplasia, pharyngeal muscle discoordination due to neuromuscular disorders [10]. Lack of
appropriate response to antibacterial substances and supportive therapy for treatment of bacterial and viral infection along with lack of maternal history of chorioamnionitis or intrauterine infection (as a source of infection), suggested that another differential diagnosis rather than pneumonia are the causes of disease and other confirmatory imaging such as CTS is necessitate for precise diagnosis.

Considering the low prevalence of neuroblastoma in the thorax of neonates, we present this case report with the first diagnosis of neonatal pneumonia and TEF and the final diagnosis of neuroblastoma.

2. Case presentation

A 17-day-old female neonate delivered by caesarian section was reported in the study with no parents’ consanguinity from a Gravid 2, Para 2, Live 2, and Abort 0 mother, at 38 weeks of gestation and birth weight of 3100 g. The mother had a normal pregnancy without any reported complications. The newborn admitted to a neonatal intensive care unit (NICU) in her hometown, because of respiratory distress, stridor, and tachypnea which was started from 7 days of life. According to parents report, intraterine ultrasonography was normal with no abnormality in the fetus and, their neonate didn’t have any problems from birth to 7 days of life.

During the first admission of the patient, based on the radiologic findings in chest x-ray in the form of aspiration pneumonia, antibiotic therapy (ampicillin and cefotaxime) and oxygen with oxyhood were ordered and due to stridor, Pulmicort and salbutamol inhaler prescribed. Unfortunately, due to the lack of recovery, the patient was discharged from the hospital with parental consent and referred to our NICU. Regard to a persistent abnormal chest x-ray of the patient despite the mentioned treatment, consisted of opacity in the right upper lobe of lung in favor of collapse consolidation and atelectasis(Fig. 1) probability of the TEF, GERD or other obstructive lesions was noticed and upper gastrointestinal tract study with thin barium was ordered which resulted in no evidence of TEF, but esophageal regurgitation during ingestion of contrast was seen and a mass like lesion in the size of 35.34 mm adjacent to upper intra-thoracic esophagus with mild mass effect was observed (Figs. 2 and 3) and thoracic CTS with contrast media was suggested for a better evaluation. Respiratory failure due to severe respiratory distress, upper airway obstruction and significant stridor in the patient on the third days of admission, led to intubation and mechanical ventilation of the neonate.

After relative stabilization of the patient, thoracic CTS was done and showed, a posterior mediastinal mass with the size of 42 × 33 × 41 mm, in right paravertebral region which crossed to the contralateral side and extending from C7-T6 area. This mass had calcification and extension to the spinal canal at the level of C7 to T4. Severe compression on the trachea and resulted in severe luminal narrowing was seen in the CTS. (Fig. 4). Based on these results neuroblastoma was suggested. Splaying and scalloping of adjacent ribs are also noted.

Due to severe obstruction on the airway and threaten of the life of the patient, the baby underwent emergency surgery by an expert pediatric surgeon. According to the operative description report, after
right posterolateral thoracotomy, a mediastinal mass with some necrotic area was seen in the superior and posterior part, adhesion and involvement of the ribs and spinal cord were observed. The resection of the tumor completely in the form of debulking was done.

In the pathological assessment of the Intra-operative biopsy was shown neuroblastoma with poorly differentiated subtype, tumor calcification, and minimal tumor necrosis. Lymphovascular invasion not identified and the diagnosis of neuroblastoma was confirmed. According to the international neuroblastoma pathology classification (INPC), favorable histopathology was reported (Figs. 5–7).

Bone marrow aspiration was shown hypocellularity in all series of erythroid, myeloid and megakaryocytic. The malignant cell was not seen in the bone marrow.

As an adjuvant laboratory test, Vanylmandelic acid (VMA) of the urine was measured with a result of 5 mg/gr Cr (it was in the normal range).

Three to four days after surgery, the patient was extubated from the mechanical ventilation and after healing the site of thoracotomy, the newborn was transferred to hematology and oncology ward of the hospital under the supervision of a qualified pediatric oncologist for chemotherapy.

3. Discussion

In this case report, we presented a female neonate with prolonged respiratory distress and stridor with a final diagnosis of neonatal
neuroblastoma. Pneumonia is a leading cause of neonatal infection and accounts for up to 10% of childhood mortality in the world especially in the developing countries [11]. Late-onset pneumonia (within the 4 and 28 days of life) caused by a variety of microorganism mainly bacteria followed by, virus, fungal and protozoa. Unfortunately in most cases, the precise diagnosis of bacterial pneumonia with detection of microorganisms in the lung tissue isn’t possible [12], and combination of clinical manifestations such as respiratory distress, apnea, tachypnea, fever and poor feeding with radiologic finding such as collapse consolidation, peribronchial thickening, confluent opacities, led to an initiation of treatment modalities. Recovery of the patient with subsiding the abnormal clinical manifestation along with the disappearance of radiologic abnormalities confirm the first diagnosis of neonatal pneumonia, otherwise, the accuracy of the diagnosis of this common neonatal infection is doubtful and other less common disorders should be considered. In this case report we present a healthy term female neonate from a normal pregnancy with no underlying risk factors for pneumonia. The patient had a persistent life-threatening respiratory distress were seen [17].

Fig. 8. Marked staining for Synaptophysin IHC marker(x20)

In a report by Nam of 17 patients with neuroblastoma, 88.2%, of the patients survive but a high ratio of stage 4S and stage 4 tumors and MYCN amplification was observed. The authors suggested that in neonatal neuroblastoma with the size of more than 3 cm, early treatment might be a better and aggressive treatment for this disease makes a better prognosis [16].

Regard to a wide distribution of involvement in neuroblastoma, there is another case report by Serdar Alan in neonatal neuroblastoma with inferior vena cava syndrome and massive hepatomegaly occupying the entire abdomen as an extremely rare disease. Venous regurgitation was seen in the skin of the abdomen and the symptoms of respiratory distress were seen [17].

We present this case report because of the low incidence of neuroblastoma particularly the thoracic form of the disease in neonatal periods with a similar appearance with neonatal pneumonia (aspiration or microbial) in the x-ray.

We suggest the clinicians; predict the diagnosis of neuroblastoma of the thorax in neonates with persistent respiratory symptoms despite usual treatments in concurrence with a mediastinal mass in an anteroposterior or lateral chest x-ray.

Conflicts of interest

None declared.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2019.100874.

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