MHA MEDIHEALTH ACADEMY

Surgery on Children

Case Report

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A rare case of primary pulmonary embryonal rhabdomyosarcoma in children

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ABSTRACT

Rhabdomyosarcoma is one of the most common soft tissue sarcomas in adolescence and young adults. Histologically, rhabdomyosarcoma has many subgroups, including embryonal, alveolar, pleomorphic, and spindle cell/sclerosing rhabdomyosarcomas. More than 50% of embryonal rhabdomyosarcomas occur in the head and neck region. The retroperitoneum and pelvis are less common sites of involvement. Primary pulmonary embryonal rhabdomyosarcoma is extremely rare. A 3-year-old girl presented with symptoms of cough and shortness of breath. A mass lesion with regular contours, approximately 3x3 cm in size, containing millimetric air densities within, and having soft tissue density was detected in the upper lobe of the right lung. According to the results of bronchoscopy and biopsy, a right thoracotomy and right upper lobectomy were performed with the diagnosis of embryonal rhabdomyosarcoma. We present a case whose pathology confirmed the diagnosis of embryonal rhabdomyosarcoma.

Keywords: Embryonal, rhabdomyosarcoma, pulmonary, computed tomography

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INTRODUCTION

Rhabdomyosarcoma is one of the most common soft tissue sarcomas in adolescents and young adults.[1,2] It accounts for an estimated 4.5% of all childhood cancers, with an incidence of six cases per one million annually.[1,3] Histologically, rhabdomyosarcoma has several subgroups, primarily embryonal, alveolar, pleomorphic, and spindle cell/sclerosing rhabdomyosarcomas.[2-5] It is usually seen in children under 15 years of age.[6] More than 50% of embryonal rhabdomyosarcomas occur in the head and neck region.[7] The retroperitoneum and pelvis are less common sites of involvement.[6,8] Primary pulmonary embryonal rhabdomyosarcoma is extremely rare.[3,5,6,8]

CASE

A 3-year-old girl presented with complaints of progressive cough and shortness of breath for 1.5 months. The patient's physical examination revealed weight loss, and laboratory findings were unremarkable. A round opacity with regular contours and approximately 3x3 cm in size was observed in the upper lobe of the right lung on the chest X-ray (Figure 1). A mass lesion with regular contours and millimetric air densities and soft tissue density was observed in the upper lobe of the right lung on chest computed tomography (CT) (Figure 2 a, b). In the patient whose differential diagnosis was considered to be hydatid cyst, *Echinococcus* IHA was negative. The lesion described in the right lung on PET CT examination had an SUVmax value of 2.8 (Figure 3). An endobronchial lesion protruded from the entrance of the upper lobe of the right lung to the trachea was observed in bronchoscopy. The pathology report diagnosed small blue round cell tumor (primarily embryonal rhabdomyosarcoma). Surgery was planned based on the findings. Right thoracotomy and right lung upper

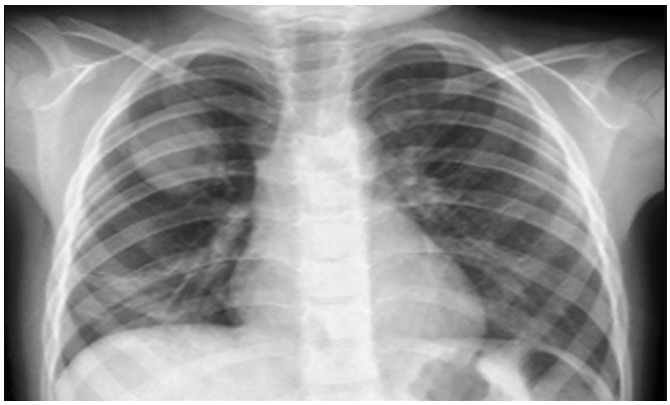

Figure 1. Chest X-ray, round opacity with regular contours in the upper lobe of the right lung

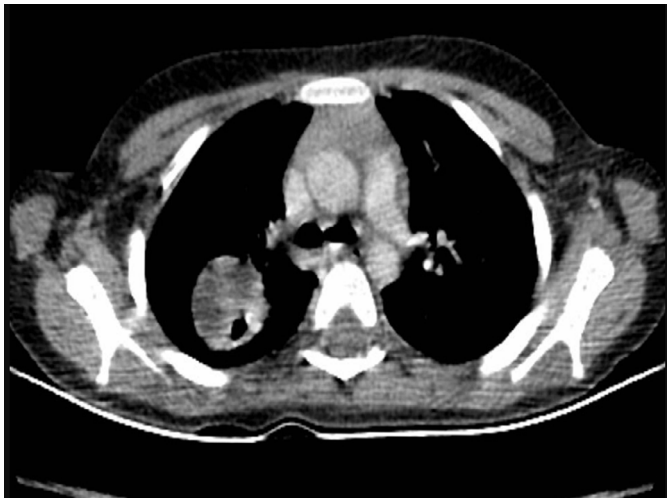

Figure 2a. Thorax computed tomography (mediastinal), a mass lesion with regular borders in the upper lobe of the right lung, containing millimetric air densities and having soft tissue density

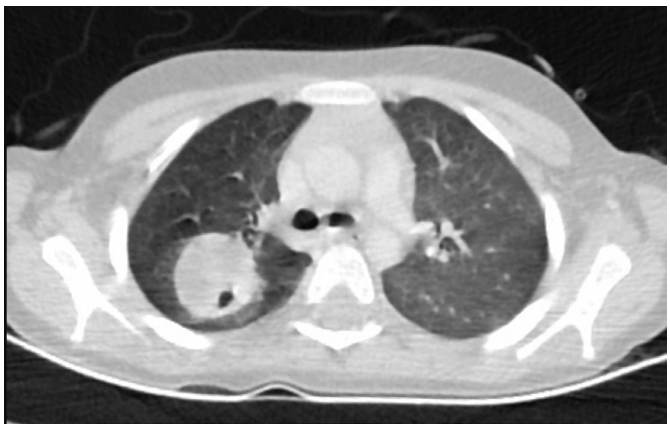

Figure 2b. Thorax computed tomography (parenchymal), a mass lesion with regular borders in the upper lobe of the right lung, containing millimetric air densities and having soft tissue density

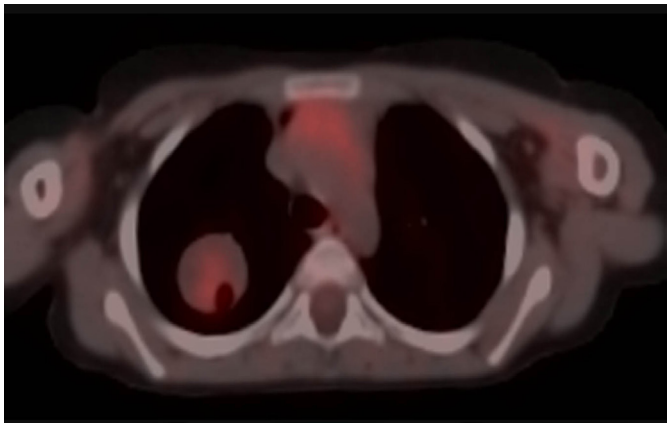

Figure 3. PET CT, a lesion with SUV max value of 2.8 in the right lung

lobectomy were performed. Pathology confirmed the diagnosis of embryonal rhabdomyosarcoma.

DISCUSSION

Rhabdomyosarcoma may develop unusually even in regions without skeletal muscle differentiation.[1,6] Primary pulmonary rhabdomyosarcoma is a very rare site for rhabdomyosarcoma to develop. Pulmonary rhabdomyosarcoma is histopathologically formed in two ways. The first is that the tumor arises from heterotrophic islands of striated muscle. The second is that the tumor arises from metaplasia of undifferentiated mesenchymal cells.[9] These tumors develop either in the context of congenital cystic lesions or from normal lung tissue. The prognosis is different in both groups. Tumors associated with congenital cystic malformations usually have a better prognosis.[9] This is because these cystic lesions are diagnosed early and are usually completely resectable lesions. On the other hand, rhabdomyosarcoma without cystic lesions presents clinically late, reaches a larger size, and cannot be completely resected.[1,6,9] There was no obvious cystic component in our case. Children with tumors smaller than 5 cm generally have a better prognosis than children presenting with larger tumors.[1,6,8-10] In our case, the tumor size was 3 cm and the tumor was completely removed surgically. Most cases present with cough, shortness of breath and chest pain.[9,10] Our case also had similar symptoms.

CONCLUSION

In the differential diagnosis of a child presenting with abnormal opacity on chest X-ray, rare conditions such as lung tumors should be kept in mind. Thus, early surgical intervention can be performed and long-term morbidity and mortality resulting from advanced stages of the disease and treatment complications can be prevented.

ETHICAL DECLARATIONS

Informed Consent

The patient's parents signed the informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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