



## CASE REPORT

### A RARE PRIMARY PULMONARY TUMOR IN CHILDREN: RHABDOMYOSARCOMA

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#### ABSTRACT

After 4 cycles of chemotherapy to treat a locally advanced tumor CT (computerized tomography) showed minimal regression of the tumor. As the locally advanced tumor was still adjacent to vital structures at the 12th week of chemotherapy, radiation therapy was given to achieve local control. The treatment was stopped after 3 additional VAC (vincristine, actinomycin D, and cyclophosphamide) courses following radiotherapy because of continued tumor progression. The patient died nine months after the diagnosis. We discussed prognostic features of primary pulmonary rhabdomyosarcoma (RMS) and recommend that it should be considered in the differential diagnosis of children with persistent pulmonary symptoms or chest X-ray abnormalities.

**Keywords:** Malignancy, Chest, Rhabdomyosarcoma, Childhood

### ÇOCUKLUK ÇAĞINDA NADİR BİR PULMONER TÜMÖR: RABDOMYOSARKOMA

#### ÖZET

Lokal olarak yaygın olan tümör, 4 kür kemoterapi sonrası çekilen bilgisayarlı tomografide minimal regresyon gösterdi. Kemoterapinin 12. haftasında halen hayati yapılara yaygınlık gösteren tümörün lokal kontrolünü sağlamak amacı ile radyoterapi uygulandı. Radyoterapiyi takiben 3 VAC (vinkristin, aktinomisin D, siklofosfamid) kürü daha verildikten sonra tümörün progresyon göstermeye devam etmesi nedeni ile tedavi sonlandırıldı. Tanıdan 9 ay sonra hasta kaybedildi. Primer pulmoner rabdomyosarkomun prognostik özellikleri tartışıldı; persistan pulmoner semptomları ve göğüs röntgeninde anormallikleri olan çocukların ayırıcı tanısında akla gelmesi gerektiği vurgulandı.

**Anahtar Kelimeler:** Malignite, Göğüs, Rabdomyosarkoma, Çocukluk

#### INTRODUCTION

Rhabdomyosarcoma (RMS), a primary mesenchymal malignant tumor with rhabdomyoblastic differentiation, is the most common soft tissue malignancy in childhood.

RMS can arise at any site, even where striated muscle is not normally present. The most common primary sites for RMS are the head and neck, the genitourinary tract, and the extremities<sup>1,2</sup>. Primary pulmonary

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rhabdomyosarcomas are extremely rare with only thirty-one pediatric cases reported in the English-language literature<sup>3-6</sup>. We report a child with primary pulmonary RMS. This case is presented for its rare occurrence in that particular location.

### CASE REPORT

A twelve-year-old female patient was admitted to the Pediatrics Ward of a local hospital with a one month history of fever, chest pain and cough. Her chest X-ray revealed complete opacity of the left hemithorax. On the 23<sup>rd</sup> day after her admission, she was referred to our hospital for evaluation of the mass and the unilateral pleural effusion in the left hemithorax that had been non-responsive to prolonged intravenous antibiotic therapy. Physical examination was within normal limits except the presence of pallor, dyspnea, and abnormal lung auscultation signs. Breath sounds were decreased in the lower zone and crepitant rales were detected in the upper zone of the left hemithorax. On chest computed tomography (CT) scan, the entire left hemithorax was found to be filled with a heterogeneous mass and unilateral left pleural effusion that was compressing and causing a total collapse of the left lung (Figure 1). All other laboratory investigations were within normal limits. No chest wall involvement was noted on magnetic resonance imaging (MRI) of the chest (Figure 2). Cytological evaluation of the pleural effusion did not reveal any malignant cells. As systemic scans and a bone marrow aspiration/biopsy failed to reveal any other primary tumor, the mass was considered as a primary intra-thoracic tumor. CT-guided trans-thoracic tru-cut biopsy specimens revealed small round cells and cross-striation. Immunohistochemically these cells stained with antibodies to the myogenic markers desmin, and myogenin. The patient was diagnosed as clinical group IIIa, stage III embryonal RMS. A VAC regimen (vincristine 1.5 mg/m, actinomycin D 1.35 mg/m, and cyclophosphamide 2.2 gr/ m) was begun according to the IRS (Intergroup Rhabdomyosarcoma Study )V protocol. The

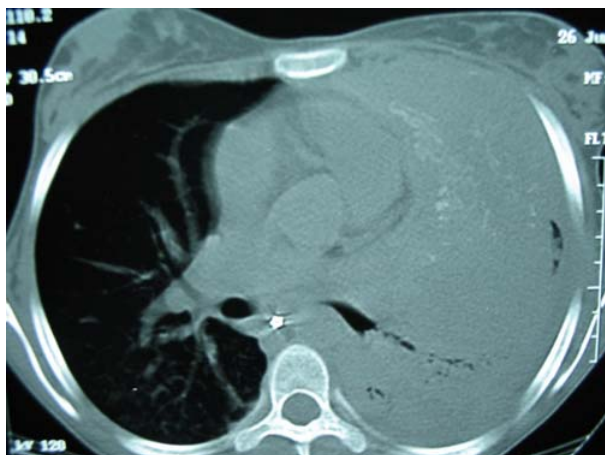
CT of the chest after 4 cycles of VAC chemotherapy revealed minimal regression of the tumor (Figure 3). As the locally advanced tumor was still adjacent to vital structures at the 12week of chemotherapy, radiation therapy was given for 6 weeks (180 cGy per day for 28 treatment days) to achieve local control. This therapy was stopped after 3 additional VAC courses following radiotherapy because of tumor progression. The patient died nine months after the diagnosis.



**Figure 1:** Axial CT scan shows a large mass on the left hemithorax



**Figure 2:** Coronal T2-weighted MRI images of the tumor on the left hemithorax



**Figure 3:** Follow-up chest CT revealed minimal regression of tumor mass on the left hemithorax

## DISCUSSION

Primary intrathoracic tumors arising from the lung in the pediatric age group are extremely rare and represent a wide spectrum of pathological conditions (pneumoblastoma, RMS, fibrosarcoma, mucoepidermoid carcinoma, pulmonary endodermal tumour and benign tumours)<sup>5,7</sup>. RMS, one of these rare pathological conditions, originates from primitive mesenchyme that has retained the capacity for striated skeletal muscle differentiation. RMS can arise at any site, even where striated muscle is not normally present, presumably from pluripotent cells that are capable of differentiating into neurogenic and myogenic elements. The head and neck are the most frequent sites of origin for RMS<sup>1,2</sup>. RMS occasionally arises in the trunk, chest wall, abdomen (including the retroperitoneum and biliary tract), and the perineal/anal region<sup>8,9</sup>. Intrathoracic region is a less common localization for RMS<sup>10-14</sup>. Primary pulmonary rhabdomyosarcomas are extremely rare and occur in a minority of patients with thoracic rhabdomyosarcomas. Prior to this case, a literature review disclosed only thirty reported cases of primary pulmonary rhabdomyosarcomas<sup>3,4</sup>. Primary pulmonary RMS can be divided into two groups: tumors in the normal lung, and tumors in congenital cystic malformation of the lung<sup>4,6,10</sup>. Tumor behavior is different in each group. Some investigators have reported

that the presence of cystic malformations can be considered as a favorable prognostic feature in pediatric patients with pulmonary RMS<sup>4</sup>. This seems to be due to early detection and complete surgical removal of the tumor associated with cystic lesions. In our case there was no pre-existing lung malformation. According to the data of the 31 reported cases with follow-up (3 months to 12 years), there were 16 patients who had associated cystic lesions. The number of disease free patients were 11/16 (68%) with associated cystic lesions and 7/15 (46%) in the group without any detectable lung cysts<sup>4-6</sup>. The site of the primary tumor is an important determinant of the prognosis. Thoracic RMS usually presents late and has become quite large by the time of diagnosis. The tumor burden at diagnosis is also a statistically significant prognostic factor. Patients with smaller tumors (<5 cm) have improved survival compared with children with larger tumors, whereas children with metastatic disease at diagnosis have the poorest prognosis<sup>3,15,16</sup>. In addition, patients with otherwise localized disease but with proven regional lymph node involvement have a poorer prognosis than patients without regional nodal involvement<sup>17,18</sup>. In our case, the big tumor burden was the leading cause of the short survival. The extent of disease following the primary surgical procedure (i.e., the clinical group) is another determinant of outcome<sup>1</sup>. In the IRS III, patients with gross residual disease after initial surgery (Clinical Group III) had a 5-year survival rate of approximately 70% compared with a greater than 90% 5-year survival rate for patients with no residual tumor after surgery (Clinical Group I)<sup>1,19</sup>. In our case, gross total or incomplete resection of the tumor was not feasible. The eventual poor prognosis was inferred by the big tumor burden, unresectability of the tumor mass and the probable absence of a preexisting pulmonary cystic malformation.

Primary pulmonary RMS, although very rare in the pediatric age group, should be considered in young patients with a pulmonary mass. Since it usually presents as a large mass at the time of diagnosis that is



adherent to adjacent vital structures, wide and complete resection of the primary tumor is less applicable. For this reason, we recommend that RMS should be considered in the differential diagnosis of children with persistent pulmonary symptoms or abnormalities on chest X-ray.

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