Case Report

Rhabdomyosarcoma mimicking as pleural effusion

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Abstract

Rhabdomyosarcoma (RMS), a malignant neoplasm of skeletal muscle origin, is the most common soft tissue sarcoma caused by infectious disease etiology, especially in nondeveloped countries. Despite being a relatively rare cancer, it accounts for approximately 40% of all recorded soft tissue sarcomas. Alveolar RMSs are seen to occur in children, while around 80% cases occur in the first three decades of life. We present here, a case of a 12-year-old child having alveolar RMS, presenting clinically and radiologically with pleural effusion.

Keywords: Alveolar rhabdomyosarcoma, thoracic neoplasm, pleural effusion

Introduction

Pleural effusion in children is usually caused by infectious disease etiology, especially in nondeveloped countries. Although thoracic neoplasms may present with pleural effusion, this is a rare finding in rhabdomyosarcoma (RMS).[1] Sometimes, an abnormal tissue or mass itself shows a picture of pleural effusion. We report a case of RMS mimicking as a pleural effusion on chest radiography.

Case Report

A child of 12 years was admitted with chief complaints of a cough (nonproductive) and fever on/off for 3 months. The child was not in distress; however, on physical examination, there was dullness and absent breath sounds in the left infra axillary, inframammary, and infrascapular regions. Abdomen was soft with, mild tenderness on applying deep pressure and no lump or organomegaly was felt. Other systemic findings were within normal limits.

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On investigation, hemoglobin was 10.9%; blood cell count was 12,300/mm^3, with a differential count of polymorphs 68%, lymphocytes 32%, eosinophils 1%, monocytes 2%, and basophils 0%. HIV, hepatitis C virus, hepatitis B surface antigen were negative, and urine routine and microscopy showed insignificant findings of pus cells 1–2/HPF and epithelial cells 2–3/HPF. Urine culture was sterile. The sputum was examined and cultured for mycobacterium tuberculosis but was negative.

The chest X-ray was done and showed left sided pleural effusion. Thoracocentesis was attempted, but pleural fluid could not be aspirated. Ultrasonography (USG) of whole abdomen and chest was performed, which showed left-sided paraspinal mass lesion with no evidence of pleural effusion.

Contrast enhanced computed tomography (CT) revealed a large soft tissue lesion with a lobulated outline in the retroperitoneum just posterior to the left kidney. The lesion measured 6.8 cm × 5.4 cm × 8.8 cm in anteroposterior, transverse, and superoinferior dimensions. The lesion showed heterogeneous enhancement on postcontrast scans with extensive areas of necrosis within it. It was closely abutting the left psoas muscle

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which was bulky at L3 level and gave a suspicion for infiltration. There was an erosion of the underlying left 12th rib and extension of the soft tissue lesion into the posterior abdominal wall. The swelling was also seen pushing the diaphragm upward from the normal level. The lesion displaced the left kidney anteriorly and was closely abutting it. However, no evidence of the involvement of the left kidney was noted. Multiple small sized retroperitoneal and mesenteric lymph nodes were seen. There was no evidence of erosion of the adjoining costovertebral junction or vertebral body. No extension into the spinal canal was noted. The appearance favored an infective process? Tuberculosis or a strong possibility of tumor [Figure 1].

USG-guided aspiration was done, and pustular fluid was aspirated which was sent for cytological and microbiological examination. Acid-fast *Bacilli* staining, Lowenstein-Jensen culture, and aerobic cultures were negative, while cytology showed only degenerative cellular debris in smears.

The patient was then subjected to tissue biopsy which showed poorly differentiated round or oval tumor cells, arranged in alveolar pattern and clusters of cells separated by fibrosis Septae (Figure 2). These clusters showed loosely placed cells in the center and tightly packed cells at the periphery. The tumor cells had N:C ratio with vesicular nucleus and prominent nucleoli [Figure 3]. Atypical mitosis was present. A diagnosis of alveolar RMS was given after confirmation with special stain: Masson's trichrome stain, which showed positivity [Figure 4].

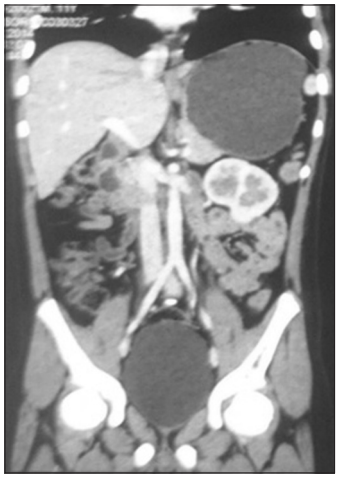

Figure 1: Coronal section of contrast-enhanced computed tomographyCECT abdomen image that revealed a retroperitoneal tumor

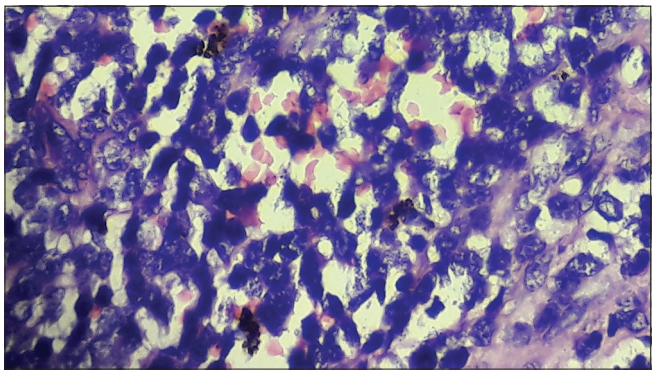

Figure 3: Alveolar rhabdomyosarcoma (H and E, ×40)

Discussion

RMS, a malignant neoplasm of skeletal muscle origin, is the most common soft tissue sarcoma, is an aggressive form of cancer developing from cells that have failed to fully differentiate.[1] It is generally considered to be a disease of childhood, as the vast majority of cases occur in those below the age of 18.[2] It is commonly described as one of the "small, round, blue cell tumors of childhood" due to its appearance on an H and E stain. Despite being a relatively rare cancer, it accounts for approximately, 40% of all recorded soft tissue sarcomas.[3]

Alveolar RMSs are seen to occur in children; Ahmad *et al.* observed over 80% cases of ARMS occurred in the first three decades of life.[4] Accurate and quick diagnosis are often difficult due to the heterogeneity of these tumors and a lack of strong genetic markers of the disease. RMSs are known to occur at any site in the body but are more commonly seen in head, neck region, genitourinary tract, or extremities.[5] Chest and abdomen are a very rare site.[6,7] Tumor presents as a swelling or mass, and many a times, the clinical features may be masked. Our case presented in the surgery department with generalized signs and

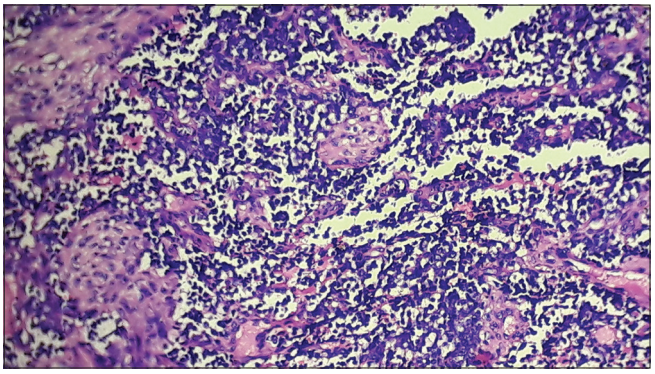

Figure 2: Alveolar rhabdomyosarcoma (H and E, ×10)

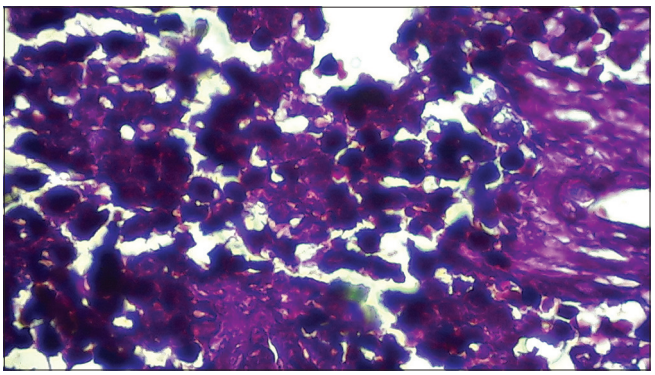

Figure 4: Masson's trichome stain stain cytoplasmic positivity

symptoms and a provisional diagnosis of psoas abscess was made. The patient was referred to us for the complaints of a cough with pleural effusion on chest radiography. After detailed clinical history and investigations, a final diagnosis of RMS (presenting as left pleural effusion) was established.[8]

Abnormal swellings have somewhat confused physicians in various ways. Lin *et al.* in his series of 14 patients stated that epithelioid angiosarcoma diffusely involving the pleural, peritoneal, or pericardial cavities, resulted in a picture closely resembling mesothelioma.[9] In our case, X-ray finding of pleural effusion was misleading and confusing, as no signs and symptoms of breathlessness were seen. This case stresses on the importance of using three-dimensional imaging techniques in a setting of misleading presentations.

Childhood cancer is a leading cause of death in children of 1–15 year's age in the USA.[10] Nearly, 4000 new cases of malignant solid tumors are diagnosed each year in children. The most common soft tissue sarcoma in the first two decades of life is RMS. There are four histological subtypes: Embryonal, botryoid, alveolar, and pleomorphic. The main localization of this malignant tumor is the head and neck (43% of primaries), while only 7% of primary tumors occur in the trunk.[6] Pleural effusion as a result of metastasis is reported to be very rare. The first case of RMS arising at the pleura was documented by Hamada *et al.* and the previous reports of RMS arising at unusual sites were also reviewed in this case report.[11]

Effective treatment of RMS requires a combination of surgery, chemotherapy, and irradiation. Surgical treatment followed by chemotherapy and radiotherapy are the methods of choice for treatment in most of the retroperitoneal tumors. Early diagnosis and multimodality treatment improves prognosis. The overall morbidity is 13% after surgical treatment of retroperitoneal tumors.[12] CT is more sensitive than chest radiography to detect abnormalities in chest as well as other part of the body. USG is considered to be very helpful for abdominal swellings or fluid in the pleural space. Histological and cytological investigations remain the ultimate diagnostic modalities.

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Conflicts of interest

There are no conflicts of interest.

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