

Mediastinal mass

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CASE

A 61-year-old Hispanic woman initially presented to the hospital with generalized weakness for two months. She had chest tightness, progressive dyspnea, and significant weight loss. A computed tomography scan (CT) of the chest showed a 9.1×7.8×10.8 cm anterior mediastinal mass with central necrosis (Figure 1). Computed tomography scans of the abdomen and pelvis were negative. Surgical resection of mediastinal mass was performed, and pathology reported a pleomorphic

rhabdomyosarcoma. A one month follow-up CT scan after surgery documented a recurrent anterior mediastinal mass (16×18×11 cm) encasing the thoracic aorta and compressing the right pulmonary artery and superior vena cava (Figures 2 and 3). The patient was referred to MD Anderson Cancer Center in Houston for an opinion about treatment; palliative chemotherapy with vincristine, cyclophosphamide, and doxorubicin was recommended. After the first cycle of chemotherapy, her clinical status deteriorated. She had difficulty breathing and hypoxemia due to the mass. She died four months after her initial diagnosis.

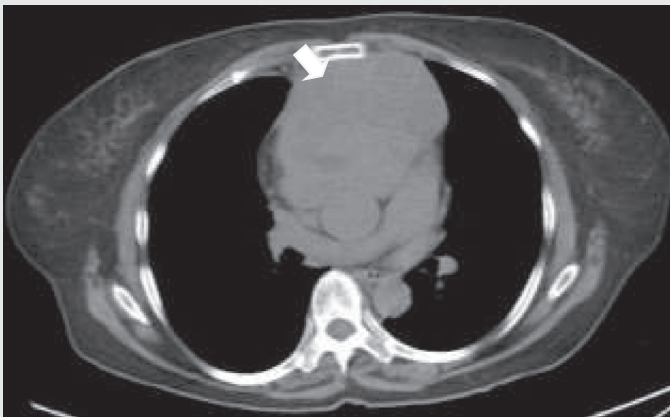


Figure 1. CT scan of chest without contrast shows a 9.1×7.8×10.8 cm anterior mediastinal mass (arrow) before surgical resection.

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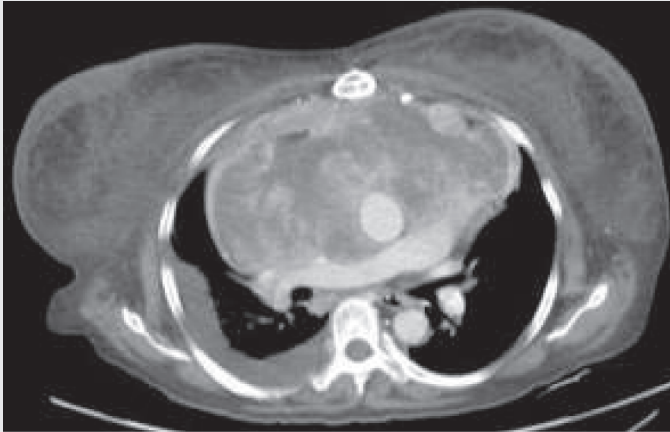


Figure 2. One month after surgical resection CT scan of chest with contrast shows a recurrent 16×18×11 cm mass which encases the ascending aorta and the pulmonary trunk.

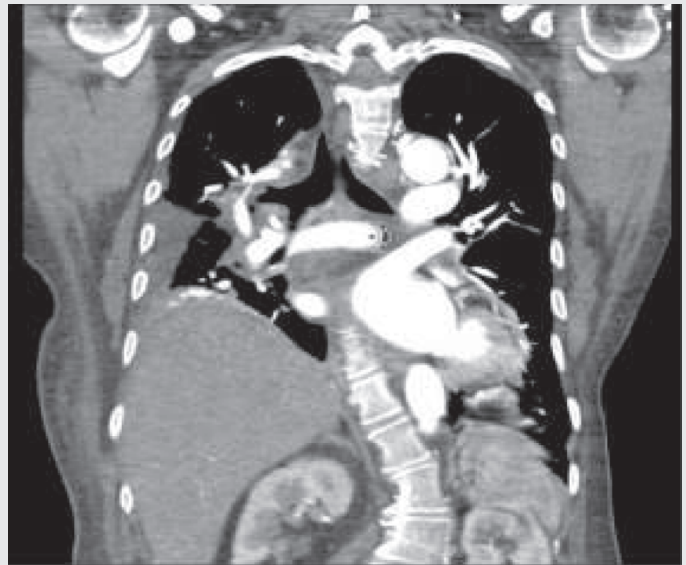
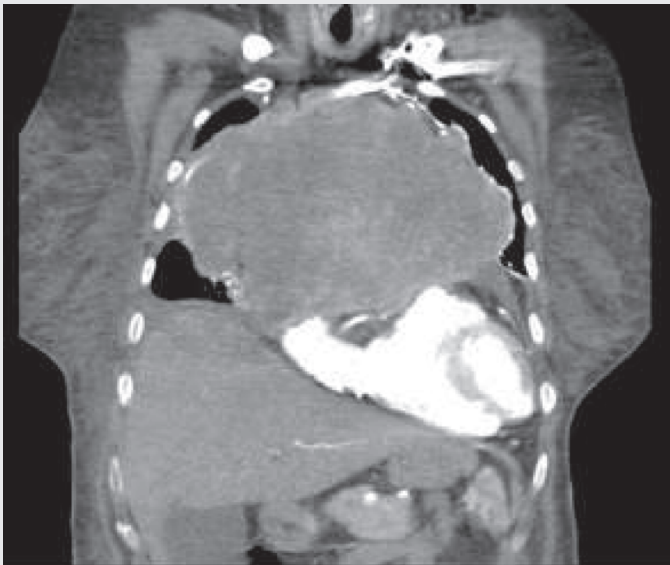


Figure 3. Coronal views of CT chest shows the mass surrounding the trachea, right and left main bronchi, and the pulmonary trunk.

DISCUSSION

Rhabdomyosarcoma is a soft tissue tumor which arises from mesenchymal cells. It is more common in children but occasionally occurs in adults. Anterior mediastinal masses are usually caused by thymomas, germ cell tumors, lymphomas, or thyroid tumors. Rhabdomyosarcoma originating from the mediastinum is very rare; these tumors usually develop in the head and neck, genitourinary tract, and extremities. A retrospective study of rhabdomyosarcoma in 76 adults aged more than 40 years old by Yu and Wang reported that 32.8% of tumors developed in the extremities, 25% in the head and neck, 21% in the trunk, and 21% in the genitourinary tract.¹ There are three major histological types of rhabdomyosarcoma: embryonal, alveolar, and pleomorphic. Embryonal and alveolar types more commonly occur in children; the pleomorphic type is the most common type in adults.² The histological type of tumor determines the primary origin of tumor and the prognosis. Embryonal type tumors have a more favorable prognosis than alveolar and pleomorphic types. Tumors that originate in the head and neck (non-parameningeal), genitourinary tract (non-bladder/prostate), and orbits have a better prognosis than tumors in the parameningeal area, prostate, bladder, and extremities.³ A pleomorphic pattern is present in 38% of these tumors, and the overall survival of pleomorphic rhabdomyosarcoma cases is eight months.⁴

Treatment modalities include surgery, chemotherapy, and radiation. Choosing the mode of treatment depends on the extent of disease, tumor location, and the histological type of tumor.

Surgical resection is usually the primary treatment except in unresectable tumors or advanced multisite disease. Brachytherapy should be considered if close or positive margins are suspected after resection. Neoadjuvant chemotherapy is used for large tumors and those involving or adjacent to vital structures to increase the chance for a surgical resection. The

most common agents used are vincristine, actinomycin, cyclophosphamide, doxorubicin, ifosfamide, and etoposide.⁵ Most patients received a multidrug regimen that includes cyclophosphamide or ifosfamide, in addition to doxorubicin, epirubicin or dactinomycin, and vincristine.⁶

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