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Image Article

A Case of Primary Pulmonary Malignant Peripheral Nerve Sheath Tumor in a Patient with Neurofibromatosis

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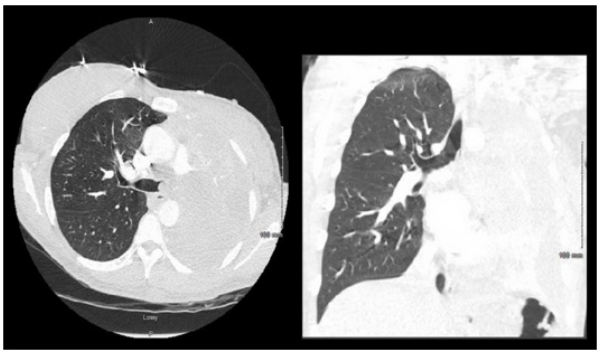
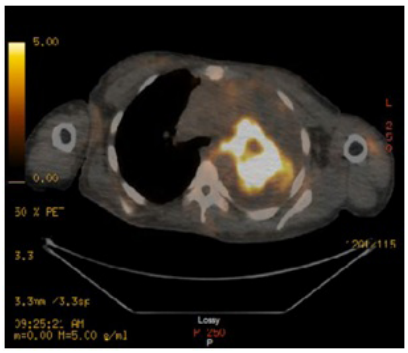
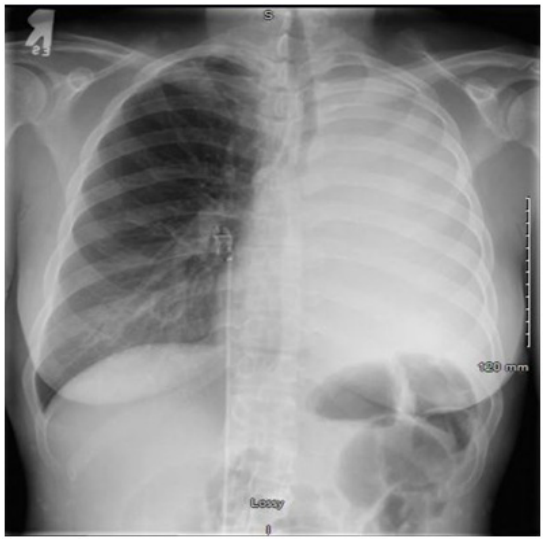
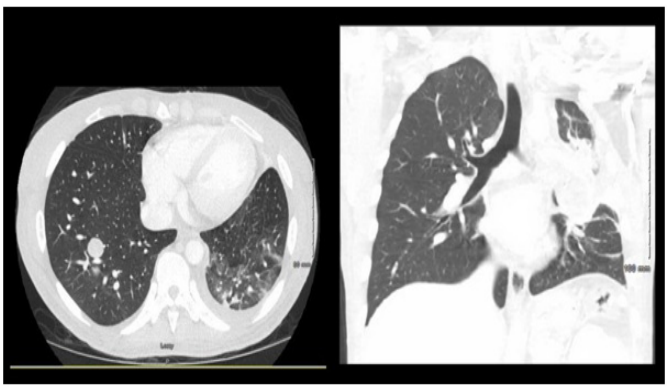
A 27-year-old lady with a history of neurofibromatosis-1 and cognitive impairment, presented with several days of progressive dyspnea. She had recently delivered a healthy baby and for some time her dyspnea had been attributed to her pregnancy. On exam, she hadinnumerable neurofibromas on face, back, trunk and extremities. Breath sounds were absent in the left hemithorax, along with dullness to percussion. Chest X ray showed complete opacification of the left hemithorax (Figure 1). CT chest showed large partially necrotic left perihilar mass with extension into the-bronchial tree, left lung collapse and left pleural effusion (Figure 2). PET scan showed the lung mass to be PET avid (Figure 3). Bronchoscopy revealed the tumor to be completely obstructing left mainstem bronchus, biopsies were obtained. Histopathology showed a hypercellular spindle cell neoplasm with immunohistochemistry confirmingmalignant peripheral nerve sheath tumor (MPNST). Patient was treated with radiation to the left hemithorax. After 5 fractions of radiation therapy, significant decrease was noted in the tumor size (Figure 4).









MPNSTs are a rare type of soft tissue sarcomas with incidence of 0.001% in the general population; however incidence substantially increases with neurofibromatosis-1 [1] Most of the cases originate in the extremities and abdomen, with lungs being an extremely unusual site for a primary MPNST. Even though MPNSTs are relatively more common with neurofibromatosis-1, almost all cases of primary lung MPNSTs have been described in patients without neurofibromatosis [2,3]. We believe our case to be one of the first reports of primary pulmonary MPNST in a patient with neurofibromatosis-1.

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