Huge intrathoracic desmoid tumor

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Abstract:
Desmoid tumors are soft-tissue neoplasms arising from fascial or musculo-aponeurotic structures. Most reported thoracic desmoid tumors originate from the chest wall. However, intrathoracic desmoid tumors are rare. We present a case of a 35-year-old male patient complaining of mild shortness of breath. The patient was diagnosed to have a huge intrathoracic desmoid tumor, which was successfully resected.

Key words:
Chest wall, desmoid, fibromatosis, intrathoracic, tumor

Discussion
John Macfarlane first described desmoid tumors in 1832. The overall incidence of desmoid tumors is approximately 2-4 cases per million of the population per year. They are rare tumors, which were given different names: Desmoid fibroma, aggressive fibromatosis, desmomas, and desmoplastic fibroma. Currently the name sarcoma of low-grade malignancy or aggressive fibromatosis is preferred due to its vulnerability to local invasion and frequent recurrences even after complete surgical resection.

They most often arise from the musculo-aponeurosis of the abdominal wall. Common sites of extra-abdominal desmoid tumors include the extremities, head and neck, and chest wall. The incidence of chest wall desmoid tumor has been reported to be 10% to 28%. Although desmoid tumors of the chest wall account for approximately 20% of all desmoid tumors, only 26 case reports...
Abnormal scarring secondary to previous surgery\cite{6,17-21} or chest trauma\cite{22,23}; hormonal factor, particularly estrogen; genetic (familial) predisposition, in relation to clonal abnormalities carried on Y chromosome, or the long arm of fifth, which is related to chromosome playing Gardner's syndrome. As many as 33\% to 38\% of patients with Gardner's syndrome develop desmoid tumors, but only 2\% of patients with desmoid tumors have Gardner's syndrome or other pathology (familial adenomatous polyposis, osteomas and other soft-tissue neoplasms)\cite{16,24-28} or abnormalities in connective tissue synthesis. Symptoms are rare and result mainly from the local mass effect of tumor encroachment on vital structures or erosion of adjacent bone or joint tissue.\cite{13}
Complete resection of the tumor with a clear surgical margin is currently the mainstay of curative treatment for desmoid tumors. The recurrence rate is high and varies from 29% to 54% in some reported series. Regular follow-up imaging is mandatory even when surgical margins are free of tumors. Re-excision is recommended for local recurrent disease. Other treatment methods, including radiation, chemotherapy, c-AMP modulation, estrogen, and prostaglandin inhibition, have been tried with varying success.

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