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Case report - Thoracic general

An uncommon tumor of the diaphragm malignant schwannoma

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Abstract

Primary neoplasms of the diaphragm are rare. Neurogenic tumors of the diaphragm, particularly those showing malignant degeneration are quite rare. We report herein a case of a 48-year-old woman, who was found to have a malignant schwannoma of the diaphragm.
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Keywords: Neurogenic tumor; Diaphragm; Malign schwannoma

1. Introduction

Primary neural tumors of the diaphragm are rare, representing approximately 10% of the neoplasms of the diaphragm. Since the first report of Klassen et al. [1], only 13 cases of diaphragmatic neural tumors have been reported up to date [2]. All of these reports comprised benign diaphragmatic schwannomas, except three cases of malignant schwannomas, which all were neurofibrosarcomas [3,4]. We are not aware of any previous report describing a malignant schwannoma of the diaphragm in the English literature.

We report herein a unique case of a rarely occurring malignant schwannoma of the diaphragm, which was successfully excised through a transthoracic approach.

2. Case report

A 48-year-old smoker woman presented with right pleuritic pain of 6-month duration. She was referred with a right-sided lung mass upon a chest X-ray. Physical examination showed dullness on percussion and decreased breath sounds in the lower zones of the right lung. Laboratory data, respiratory function tests and arterial blood gas analysis were within the normal limits. Chest X-ray showed right-sided non-homogeneous mass in the lower field of the lung. Computed tomography (CT) revealed a posteriorly located mass in the right hemithorax measuring 15 × 10 cm in diameter, and in close association with the diaphragm (Fig. 1).

Abdomen utrasonography revealed that the tumor did not originate from the liver. Bronchoscopy was normal.

On right posterolateral thoracotomy, a huge, lobulated and well-demarcated mass, located in the posterior mediastinum, arising from the diaphragm was explored. No evidence of invasion to the lung parenchyma or adjacent structures was observed. The tumor was excised including a part of the diaphragm measuring 4 × 5 cm in diameter and the diaphragm was repaired primarily.

The entire tumor was measured 33 × 30 × 17 cm in diameter and weighed 1520 g. On cut section the mass was soft and pale yellow in color. On histologic examination, tumor consisted of loosely arranged fibroblast-like indifferentiated spindle cells with focal lipoma-like areas. Spindle cells showed mild to moderate pleomorphism with rare mitosis some being atypical. A maze of branching narrow vessels was distinct in some areas. Focal areas of osseous metaplasia were also observed. Immunohistochemical study for cytokeratin, SMA, CD68, FVIII, CD34, and desmin was negative in the tumor cells. Vimentin was positive and S-100 stained the lipoma-like areas. Spindle cells only showed vimentin expression. The tumor was diagnosed as a malignant schwannoma (Fig. 2). Retrospective histological examination of the formerly excised tumor revealed that the present tumor was the recurrence of this tumor.

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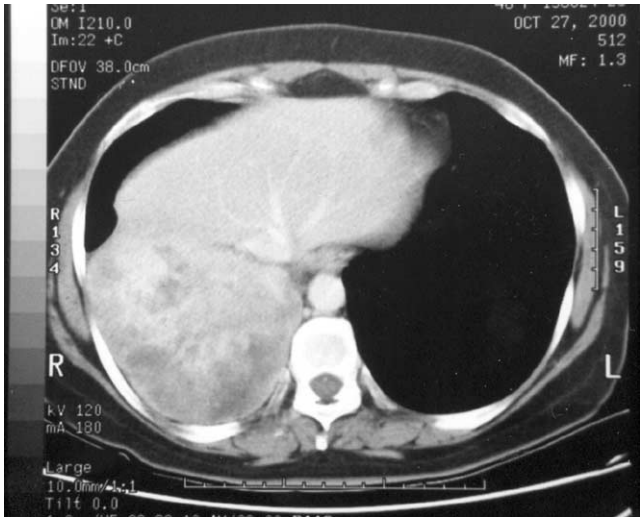


Fig. 1. Thorax CT showing posteriorly located mass in the right hemithorax.

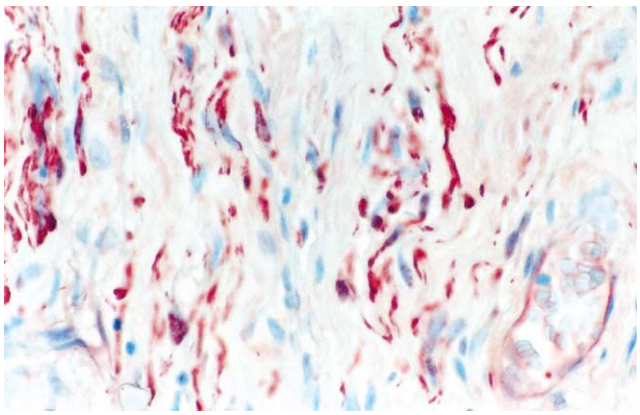


Fig. 2. Short fascicles of schwann cells showing strong immunoreactivity with S-100 protein (streptavidin biotine peroxidase ×400).

Postoperative course was uneventful. The patient remains well and disease-free 32 months after surgery.

3. Discussion

Grancher was the first to discover the primary tumor of the diaphragm during an autopsy study [5]. Wiener and Chou [6], who documented 71 diaphragmatic tumors, reported the larger series for the first time. Since then Olafsson et al. [7] reported 14 additional cases. Of these 85 total cases, 33 (39%) comprised the malignant tumors of the diaphragm. McClenethan [2] reported a case of a primary neurilemmoma of the diaphragm with a review of the diaphragmatic tumors with neural origin. The review revealed three cases of malignant tumors arising neural tissue, which all were neurofibrosarcomas. Thus, our case is unique, in that the tumor of the diaphragm was a malignant schwannoma.

Patients with diaphragmatic tumors often present between 40 and 60 years old. Men and women are affected equally. Most patients are symptomatic, with chest wall or abdominal pain, cough, and dyspnea as the most encountered symptoms. However, 20% of the patients are discovered incidentally on routine chest X-rays, arthralgias and hypertrophic pulmonary osteoarthropathy presenting as clubbing are rare manifestations, which may resolve following excision of the tumor [2,6,8]. Our patient was also symptomatic, and had pleuritic pain.

Gastrointestinal barium studies and pneumoperitoneum have been performed in the diagnostic work-up of the diaphragmatic tumors previously. These imaging techniques have been replaced by CT and magnetic resonance imaging (MRI) currently. Ultrasonography may also be an aid in the differential diagnosis with liver and spleen lesions.

The treatment of choice for diaphragmatic tumors is surgical excision. Although laparatomy with subcostal incision has been reported as an alternative for surgical approach of the diaphragmatic tumors [8], thoracotomy will provide an optimal exposure of the involved diaphragm [2,9].

References

[1] Klassen KP, Patton R, Bemen FM. Neurofibroma of the diaphragm. J Thorac Surg 1945;14:407–13.
[2] McClenathan JH, Okada F. Primary neurilemmoma of the diaphragm. Ann Thorac Surg 1989;48:126–8.
[3] Samson PC, Childress ME. Primary neurofibrosarcoma of the diaphragm: report of two cases. J Thorac Surg 1950;20:901–10.
[4] Doyle T. Left basal opacity for diagnosis. Chest 1983;84:199–200.
[5] Grancher M. Extraits des proces-verbaux, 8. Tumeur du diaphragme. Bull Soc Anat Paris 1868;43:385.
[6] Wiener MF, Chou WH. Primary tumors of the diaphragm. Arch Surg 1965;90:143–52.
[7] Olafsson G, Pausing A, Holen 0. Primary tumors of the diaphragm. Chest 1971;59:568–70.
[8] McHenry CR, Pickleman J, Winters G, Flisak ME. Diaphragmatic neurilemmoma. J Surg Oncol 1988;37:198–200.
[9] Moinuddeen K, Baltzer JW, Zama N. Diaphragmatic eventration. An uncommon presentation of a phrenic nerve schwannoma. Chest 2001; 119:1615–6.

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