Case report
Primary mediastinal malignant meningioma

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Abstract
Primary ectopic meningiomas are extremely rare tumors of controversial origin and they are usually limited to the head and neck region. Its occurrence at the mediastinum is even rarer. There has not been any official report regarding primary mediastinal malignant meningioma until today. Because of its rarity and potential value, we report here a case of primary mediastinal malignant meningioma, which turns out to be the first reported case of this type of meningioma. The clinical features, treatment plans, pathological findings, as well as prognosis of a case of primary mediastinal malignant meningioma were carefully analyzed and the literature on ectopic meningioma was reviewed. The diagnosis of ectopic meningioma can only be established based on microscopic and immunohistochemical findings. Surgery is the treatment of choice for ectopic meningioma and postoperative radiotherapy should be managed for patients with suspected invasive meningioma.

Keywords: Ectopic meningioma; Mediastinum; Malignant

1. Introduction
Meningiomas are common, usually benign, slow-proliferating tumors of the central nervous system (CNS), accounting for about 15—20% of all intracranial tumors. Primary extracranial and extraspinal meningiomas are rare. They have usually been found to arise in the head and neck region [1—3]; and less frequently in the regions including the foot, the lung, the skin, and the paravertebral regions [4—6]. Occurrence at the mediastinum is even rarer and only a few cases have been described in the literature until now. There are no statistical reports regarding ectopic malignant meningiomas. After all, they are assumed as ‘extremely rare’. The following case reported will present a first glance into primary mediastinal malignant meningioma.

2. Case description

2.1. Clinical features and treatment
On April 9th 2008, a 41-year-old man was admitted to our hospital suffering from progressive chest distress and coughs, which had lasted for about 1 month. On admission, his general condition was stable. His thorax was proved to be normal after physical examination, but chest roentgenogram revealed an oval-shaped mediastinal mass in the right hemithorax. Computed tomography (CT) scan of the chest revealed a huge mass of soft tissue density at his right anterior mediastinal compartment (Fig. 1). The mass was about 10 cm in its greatest dimension and located anteriorly on the right lateral side of the aorta. Its border adjacent to the superior vena cava was poorly demarcated. The mass was of non-uniform density accompanied with areas of necrosis after contrast enhancement. The tumor was carefully excised and was found to have infiltrated the superior and middle lobe of the right lung, as well as the wall of superior vena cava. Dysplasia of the right superior lobe was significant. Based on these surgical findings and subsequent consents obtained from patient's family members, superior lobectomy was performed. The whole surgery process went smoothly. A small residual tumor tissue was left untouched at the superior vena cava wall and it was marked. Histopathological examination showed evidences of malignant meningioma. Following the diagnosis, the patient was underwent a postoperative magnetic resonance imaging (MRI) of the head and spine, which appeared to be normal.
The patient received a course of radiotherapy one month after surgery and showed no signs of any recurrences or metastasis during a 7-months follow-up.

2.2. Pathological findings

The lesion showed uniform spindle cell proliferation separated by collagen bundles (Fig. 2). Abundant eosinophilic cytoplasms were seen and nucleus was lightly stained. Infiltration of tumor cells was seen in the resected lung tissues. Small necrotic foci were seen in tumor tissues. Immunohistochemical study was used to identify the nature of the tumor with standard avidin–biotin. The neoplastic cells were revealed to be positive for epithelial membrane antigen (EMA), CD34, CK-P, Ki-67 and vimentin and negative for CD99 and bcl-2. Its morphologic and immunohistochemical findings were compatible with the diagnosis of meningioma.

3. Discussion

Primary extracranial and extraspinal meningiomas are exceedingly rare and have been reported sporadically. The histogenesis of ectopic meningiomas was not entirely clear. It is believed that these tumors are derived from ectopic arachnoid cell [7] or from mesenchymal cells or Schwann cells, which have been differentiated into meningotheelial cells [8].

It is difficult to make a definite diagnosis of ectopic meningioma and it can only be established based on microscopic and immunohistochemical findings. In this case, despite the unusual location of the tumor, its pathological findings closely resembled those of a CNS meningioma. Surgical and pathological findings further disclosed the anatomical relationship between the tumor and the spinal cord more clearly and there were also no clinical and radiological evidences of any intracranial or intraspinal lesion. These have shown that the meningioma in this case is of complete mediastinal origin. According to our reviews on the literature, this is the first reported case of primary mediastinal malignant meningioma.

In summary, although primary mediastinal malignant meningiomas are extremely rare, surgical excision proves to be curative and the prognosis is also sound just as in the case described in this paper. Adjuvant radiotherapy would be necessary if a tumor is suspected to be an invasive meningioma. In the above-mentioned case, there are no signs of any recurrences or metastasis during a 7-month follow-up. With regard to the malignancy features of this tumor, we recommend that close long-term follow-ups with routine CT scans should be arranged in any of these cases.

References