Neuroradiology

Metastatic biphasic pleural mesothelioma presenting with cauda equina syndrome

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ABSTRACT

Patient with previous asbestos exposure on a watchful wait and watch regime presents acutely with cauda equina syndrome. Radiological imaging confirmed a mass with direct invasion of the spinal cord. Histology confirmed metastatic pleural mesothelioma.

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Introduction

This is a case study of a patient presenting clinically with cauda equina syndrome. After detailed imaging of the spine, it shows an intramedullary mass in the spine responsible for the patient symptoms. Tissue confirmation postbiopsy confirmed biphasic pleural mesothelioma.

Case report

An 84-year-old male plumber had previously known asbestos exposure with pleural changes managed clinically by watchful waiting since 2010 with serial computed tomography (CT) scans from 2010 until 2016. He was admitted as an inpatient in June 2016 with a 12-week history of reduced sensation in
his left leg, back pain, reduced sensation opening his bowels, difficulty walking, and reduced proprioception in his leg.

The repeat CT thorax as an inpatient (Fig. 1) showed significant new irregular pleural thickening, particularly in the left side posteriorly in the upper dorsal left chest with extension towards the left nerve root at T2/3 level with a visible enhancing mass compared with normal CT findings in 2015 (Fig. 2). There was also evidence of new disease in right lower chest with local bony erosion secondary to enhancing mass (Figs. 3 and 4).

The patient’s scans were consequently discussed in the lung cancer multidisciplinary meeting under working diagnosis of pleural mesothelioma with intramedullary compression of the spinal cord (Fig. 5, Fig. 6).

As per multidisciplinary meeting outcome, he was started on high dose dexamethasone and discussed with the local neurosurgical team. The neurosurgeons felt that surgical intervention would be more likely to cause damage to the spinal cord. He was offered palliative radiotherapy to the spinal cord at this stage but he declined, preferring to await histological confirmation.

The biopsy later confirmed the mass to be consistent with biphasic pleural mesothelioma (Fig. 7, Fig. 8).

Our patient agreed for radiotherapy (20 Grays in 5 fractions of proven therapeutic effect) postbiopsy result and clinical worsening of neurology in lower limbs.

The patient was referred to palliative care team to optimize pain and comfort care due to deteriorating clinical condition. The patient died shortly after due to respiratory failure.

Discussion

Two thousand seven hundred people are diagnosed with pleural mesothelioma in rare neoplasm of the mesothelial layer of the pleura per year in the UK with survival rate of 7.5% over 5 years.
The highest incidence rates reported from Europe are the United Kingdom, The Netherlands, Malta, Belgium, Australia, and New Zealand.

Prognosis of pleural mesothelioma is generally poor from the moment of diagnosis however similar to other forms of malignancy, the mortality rates vary moderately depending on the stage at which the diagnosis is made. Stage 1 shows the median life expectancy of 21 months whereas diagnosis at stage 4 showing the life expectancy of 12 months. Survival rate percentage after diagnosis also reflects an unfavorable outcome with 38% after 1 year. This steeply drops to 10% after 5 years and 4% after 10 years.

With respect to this case study, the site and type of metastasis remain the noteworthy objective. The most common sites of metastatic spread for pleural mesothelioma are regional lymph nodes, lung, liver, adrenal glands, and kidneys. The lowest rate of the extrathoracic site of metastasis is found in the central nervous system (as low as 3%).

Histological subtype of pleural mesothelioma does affect treatment, aggressiveness of a tumor and response rates. Epitheloid variants of pleural mesothelioma have a better response than sarcomatoid cells. Therefore in a biphasic mesothelioma, if the number of epithelioid cells is high then treatment and prognosis is better for the patient.

In radioresistant tumors or rapidly declining neurological functional status, surgical intervention remains the preferred treatment option.

Radiotherapy with combination chemotherapy (doxorubicin, irinotecan, and cisplatin) has also not really shown to prolong patient survival with median patient survival being less than 1 year from the time of system onset. Most often patients with malignant mesothelioma often die of respiratory failure as is evident in the case presented here.

Given the patient clinical presentation and history in this case study, the differential diagnosis was limited to primary or secondary pleural neoplasm metastatic to the spinal cord with a remote possibility of pleural or spinal lymphoma in the differential diagnosis.

Spinal cord and brain involvement of metastatic malignant mesothelioma are not common with intramedullary metastasis (mentioned in this case study) being even rare. The mechanism for intramedullary and brain metastasis has been hypothesized to be due to hematogenous or leptomeningeal spread without direct continuity of the primary pleural lesion. Spread to the dural layer is likely due to a perineural or direct extension of the pleural neoplasm. Only 12 cases of intramedullary compression of malignant mesothelioma have been reported so far.

Intradural, extramedullary metastasis to the spinal cord and brain is often considered at a lower threshold by surgeons for resection depending on the functional and comorbidity status of the patient. However intramedullary metastatic site to the spinal cord is rarely considered for surgical decompression with beneficial results. Its identification and early clinical diagnosis is more tailored toward earlier palliative treatment and radiotherapy for symptoms control for the patient.

This case highlights the importance of clinicians being open-minded in day to day clinical review of considering metastatic pleural mesothelioma as a differential of a primary cord lesion especially if the clinical history and occupational risk factor profile favors the presentation of malignant mesothelioma.

**Conclusion**

Extrathoracic metastatic sites with pleural mesothelioma are rare. This case illustrates the importance of clinical correlation with radiological and tissue confirmation. This leads to early intervention and improving quality of life for the patient.
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Fig. 7 – Lung (arrow) with a tumor in pleura 4×. Pleura with epithelioid and necrosis (star) 10× (block arrow) and areas. Biopsy taken under ultrasound from pleural mass tapering into left T3/T4 lateral foramen showed fragments of thickened fibrotic pleura, with a biphasic tumor composed of epithelioid and spindle cells.

Fig. 8 – Epithelioid cells with mitosis (arrow) 40×. Epithelial membrane antigen immunostain positive 20×. Immunostaining confirmed a biphasic malignant mesothelioma, with the tumor cells positive for MNF116 and epithelial membrane antigen with focal positivity for CK 5/6 and few cells positive for WT-1. The latter 2 being mesothelial markers. Tumor was negative for p63, calretinin, desmin, CD34, and TTF-1. Mib-1 showed a proliferative index of about 30%.