Primary Malignant Melanoma of the Pleura: A Rare Case

Low Qin Jian[1], Benjamin Ng Han Sim[2], Cheo Seng Wee[3]
[1]Medical Department, Melaka General Hospital, Melaka, Malaysia
[2]Department of Medicine, Segamat Hospital, Johor, Malaysia
[3]Medical Department, Hospital Queen Elizabeth, Kota Kinabalu, Sabah, Malaysia
[*]Corresponding author's email: peterlow4964@gmail.com
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ABSTRACT

Primary pleural melanoma is a very rare condition and highly aggressive tumour. A patient presented with productive cough, haemoptysis, pluritic chest pain and breathlessness. On investigation, she was diagnosed as left-sided lung mass with pleural effusion. Pleural biopsy confirmed malignant melanoma of pleura and she was subsequently referred to the oncology team for palliative chemotherapy. In conclusion, primary pleural melanoma remains a rare disease with no proven effective treatment regime available.

Keywords: primary, malignant melanoma, pleural

INTRODUCTION

Malignant melanoma (MM) commonly arises from the deeper layers of the skin or the eyes and is the leading cause of death.[1] Primary malignant melanoma is a rare condition. Malignant melanoma can involve any mucosal regions such as the oral mucosa, oesophagus, larynx and the ano-genital mucosa.[1] It is commonly metastasize from skin cancers.

Less than 30 cases that have been reported in medical literature fulfilled all the requested criteria to diagnose primary pleural melanoma[2]. The proposed criteria to diagnose pleural melanoma is difficult to fulfil as all extra pulmonary origin of the tumour needs to be excluded first[3]. Unfortunately, many patients are very anxious when this diagnosis is being informed to them and hence further invasive investigation to rule out other primary sources are hard to be performed just like in this case that we encountered.

CASE PRESENTATION

A 41-year-old non-smoker Malay lady presented with two weeks history of productive cough associated with haemoptysis and pleuritic chest pain. She also reported weight loss and poor appetite during this period.

Clinically, she was breathless. Respiratory system examination was consistent with a left-sided pleural effusion. There was no palpable lymphadenopathy. Skin examination of the total skin surface revealed no melanoma. She was normotensive and non-diabetic.

Chest radiograph showed homogenous opacity in the left lung. Bedside ultrasound scan of her left lung showed a lung mass with pleural effusion. We proceeded with left pleural biopsy and a thoracostomy tube was inserted and pleural fluid drained was sent for analysis. The results came back as exudative pleural effusion based on Light's criteria. The diagnosis of malignant melanoma of the lung was confirmed by the immunohistochemistry and histomorphological report. On immunohistochemical examination, the tumour cells were positive for the expression of intracellular melan-A, human melanoma-45 (HMB-45), vimentin and S-100. It was negative for calretinin and pancytokeratin. Her positron emission tomography/computed tomography showed a left pleural mass with large pleural effusion in the left hemithorax with raised metabolic activity seen in left pleura, right lung and ribs. There was possible right lung and skeletal metastasis. Retinal examination under slit lamp did not reveal any evidence of melanoma. We had counselled her for an endoscopic examination to rule out any gastrointestinal tract source of her melanoma

but she refused. We did not investigate for leptomeninges melanoma metastasis in view that she was asymptomatic with no headache or signs of raised intracranial pressure. She was referred to the oncology team who counselled her for palliative chemotherapy. She received a cycle of chemotherapy with dacarbazine (200mg/m2, days 1 – 3) and cisplatin (30mg/m2, days 5 – 7) during her inpatient stay but subsequently took self-discharged against medical advice to seek alternative medicine opinion searching for a cure to her illness. She eventually presented after three months with severe dyspnoea and succumbed to the disease.

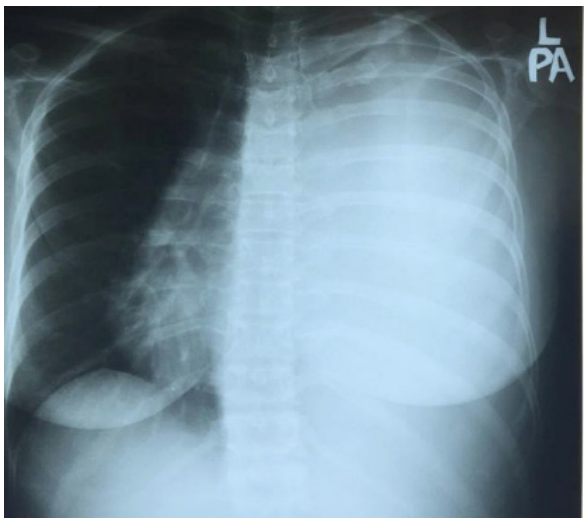

Figure 1 Chest X-ray showed homogenous opacity over the left hemithorax

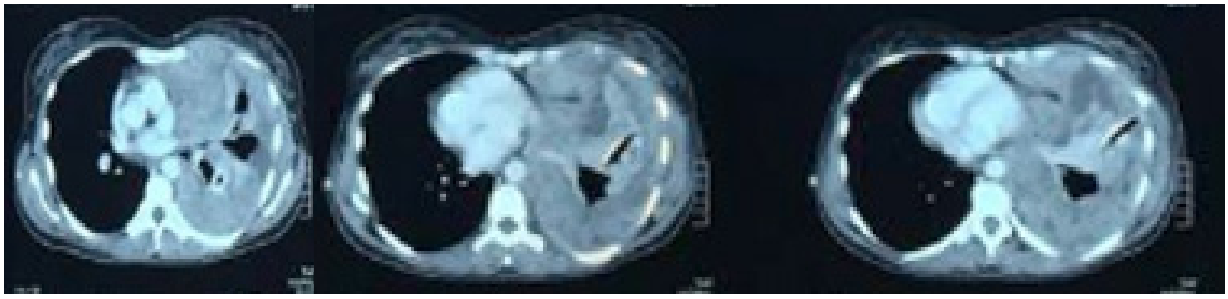

Figure 2 Computed tomography of thorax showed left pleural mass with effusion

DISCUSSION

Primary malignant melanoma (MM) of the pleural is a rare condition accounting in about 0.01% of all lung malignancy.[1] It can present endobronchially and manifest with respiratory symptoms such as cough, haemoptysis, lung collapsed or atelectasis.[3]

This condition can mimic other lung malignancy and hence a histopathological study would be beneficial. The final diagnosis of primary MM of the lung is established based on clinical, radiological and pathological findings.[3]

The proposed criteria for the diagnosis of primary MM includes the following[3]:

1. Junctional changes like 'dropping off' or nesting of the melanoma cells just beneath the bronchial epithelium.
2. Invasion of the bronchial epithelium by melanoma cells.
3. Malignant melanoma associated with these epithelial changes.
4. A solitary lung tumour.
5. No history of cutaneous, mucous membrane or ocular melanoma.
6. Absence of other detectable tumour at the time of diagnosis.

The pathogenesis of primary MM of the lung is still poorly understood.[3] One hypothesis is that melanocytes exist throughout the body as cells of a dispersed neuroendocrine system.[1] Normally, melanocytes migrate to the epidermis and the dermoepidermal junction of the skin, sometimes they can also migrate to the visceral during embryogenesis.[3] This has been suggested for the oesophagus and larynx and may be the case for the lung. The residual primitive melanoblasts which share a common origin with other melanoblasts located in the trachea, oesophagus, and pharynx, giving rise to MM of the lung. Their origin from the neuroectoderm is also said to be the cause of their low incidence in the endodermal epithelium. There is another theory that believes that melanoma cells may have been derived from pluripotent stem cells.[3]

Treatment of choice would be surgical resection.[3] The role of adjuvant chemotherapy post-operative is not known. Radiotherapy had been tried in mucosal melanoma of the head and neck with disappointing results.[1] As in this case, chemotherapy is used mainly for palliative only. The prognosis is rather poor but existing data is inadequate to conclude with conviction.[3]

CONCLUSION

Primary malignant melanoma of the lung is a rare pathological entity. It can be diagnosed with careful assessment of both clinical and histopathological studies to establish the diagnosis.

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CONFLICT OF INTEREST

The authors declare that they have no competing interests.

CONSENTS

Written informed consent was obtained from the patient to publish the case with its related pictures. A copy of the written consent is available for review by the Chief Editor of this journal.

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