VOLUME 16, NUMBER 6 NOVEMBER/DECEMBER 2007 ISSN 1054-8807

CARDIOVASCULAR PATHOLOGY

A Journal of Basic, Clinical and Applied Cardiovascular Science

The Official Journal of the Society for Cardiovascular Pathology

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1054-8807(200711/12)16:6;1-D

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Cardiovascular Pathology 16 (2007) 351–353

CARDIOVASCULAR PATHOLOGY

Case Report

Primitive neuroectodermal tumor of the lung with pericardial extension: a case report

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Received 19 December 2006; received in revised form 29 December 2006; accepted 1 February 2007

Abstract

A 16-year-old student presented with a 4-week history of progressive shortness of breath, loss of appetite, and occasional blood-tinged sputum. The chest X-ray revealed massive right-sided pleural effusion with cardiomegaly. An echocardiogram revealed a large pericardial mass with massive pericardial effusion. Subsequent computed tomography of the thorax revealed a large heterogeneous mass in the right lung with extension into the pericardium. Lung biopsy revealed primitive neuroectodermal tumor (PNET) with small round blue cells, Homer–Wright rosettes, and CD99 positivity. We discuss pericardial metastases of PNET and its implication in this patient.

Keywords: Primitive neuroectodermal tumor (PNET); Pericardium; Prognosis

1. Case report

A 16-year-old student with a history of childhood asthma presented with a 4-week history of progressively worsening shortness of breath and occasional blood-tinged sputum. Empiric treatment for an upper respiratory tract infection failed to improve his symptoms. Clinical examination revealed clinical signs consistent with a massive right-sided pleural effusion.

Electrocardiogram showed low voltage and pericarditis. Trans-esophageal echocardiogram revealed a large homogenous mass in the mid to lower mediastinum, posterior to the heart and compressing the left atrium with massive pericardial effusion (Fig. 1). Pericardiocentesis yielded 1.7 l of bloody fluid, containing only red blood cells and neutrophils on cytological examination.

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1054-8807/07/$ – see front matter © 2007 Published by Elsevier Inc.
doi:10.1016/j.carpath.2007.02.002

Computed tomography (CT) scan of the chest showed a heterogeneously enhancing mass measuring 14.7×13.1 cm (AP×Wd), occupying most of the right lower thoracic cavity and appears continuous with the pericardial lining of the right heart border and extends into the pericardial space compressing the left atrium (Fig. 2). Bone scan showed metastatic deposits to the right humeral shaft and the ninth rib.

A trans-thoracic biopsy revealed primitive neuroectodermal tumor with small round blue cells, rosette formation, and positive CD99 staining (Fig. 3). Following confirmation of the diagnosis, this patient was deemed to be of highest risk (Group 3, Stage 4) and commenced on six cycles of chemotherapy consisting of vincristine, ifosfomide, doxorubicin, and etoposide (VIDE).

2. Discussion

Primitive neuroectodermal tumor (PNET) is part of the Ewing's sarcoma family of tumors (ESFT) [1]. Primitive neuroectodermal tumors are malignant tumors of small

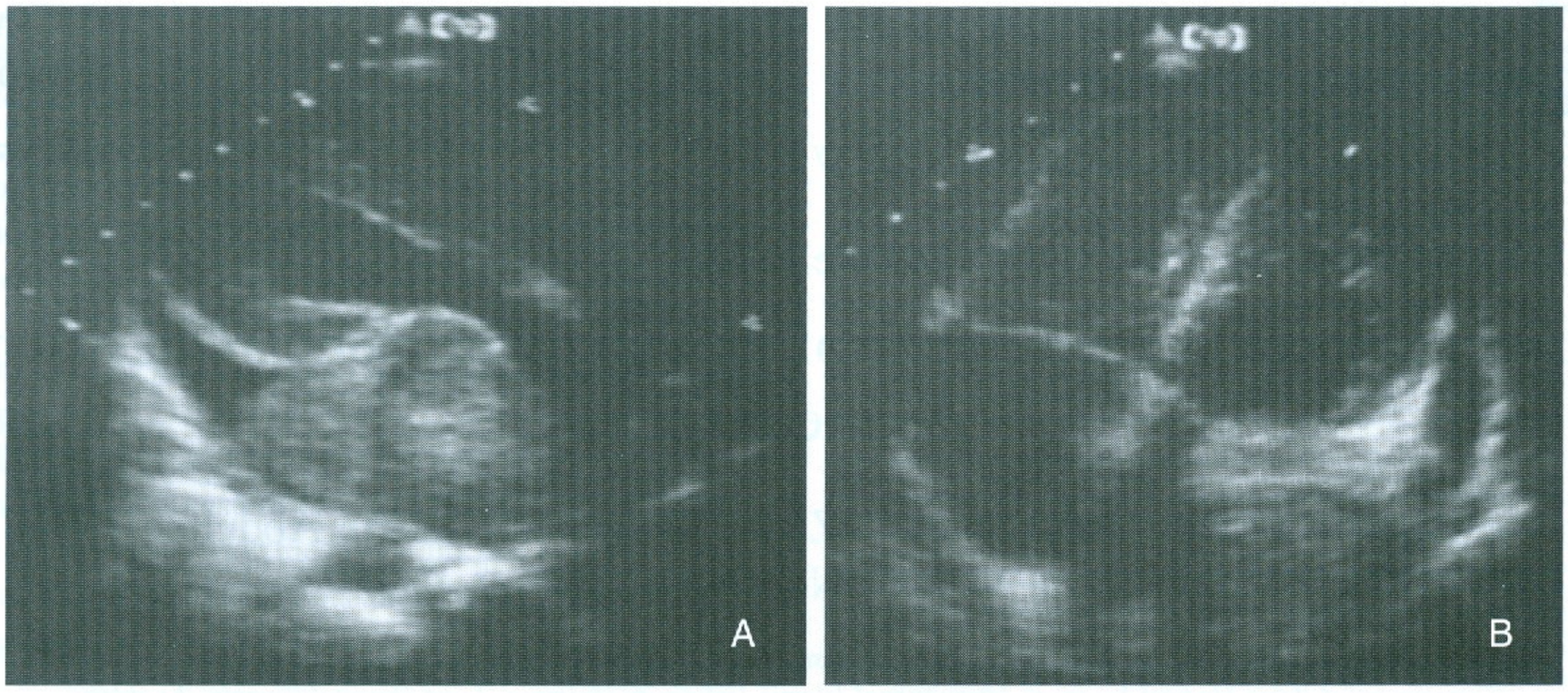

Fig. 1. Large intrapericardial mass compressing on the left atrium in the left sternal edge (A) long-axis view and (B) apical four-chamber view.

undifferentiated neuroectodermal cells thought to originate from neural crest cells and may develop in soft tissue [2]. Lung tumor with metastases to the heart is common and constitutes about one third of all metastatic heart tumors [3,4]. However, PNET of the lungs with direct extension to the pericardium is exceedingly rare, and to our knowledge, pericardial PNET has only ever been reported once [2].

Primitive neuroectodermal tumor is a highly differentiated form of ESFT and in this case is confirmed by the presence of Homer–Wright rosettes and CD99 positivity. This differentiation is essential as it carries a significant prognostic impact, being poorer for PNET [2]. This patient was also classified as being high-risk according to the EURO-EWING-99 trial and was commenced on six cycles of chemotherapy consisting of VIDE [1].

Following chemotherapy, there was regression in the size of the tumor from 14.7×13.1×12.5 cm=2407.1 cm^3 to 9.0×7.2×4.0 cm=259.2 cm^3 on thorax CT measurement. However, despite the absence of bone marrow metastases, and regression in the metastatic deposits to the humerus and ninth rib, the overall survival rate is still poor at less than 10% [5].

Acknowledgments

We would like to acknowledge the contributions of the Department of Pathology and Radiology, National University Malaysia, in providing the figures for publication.

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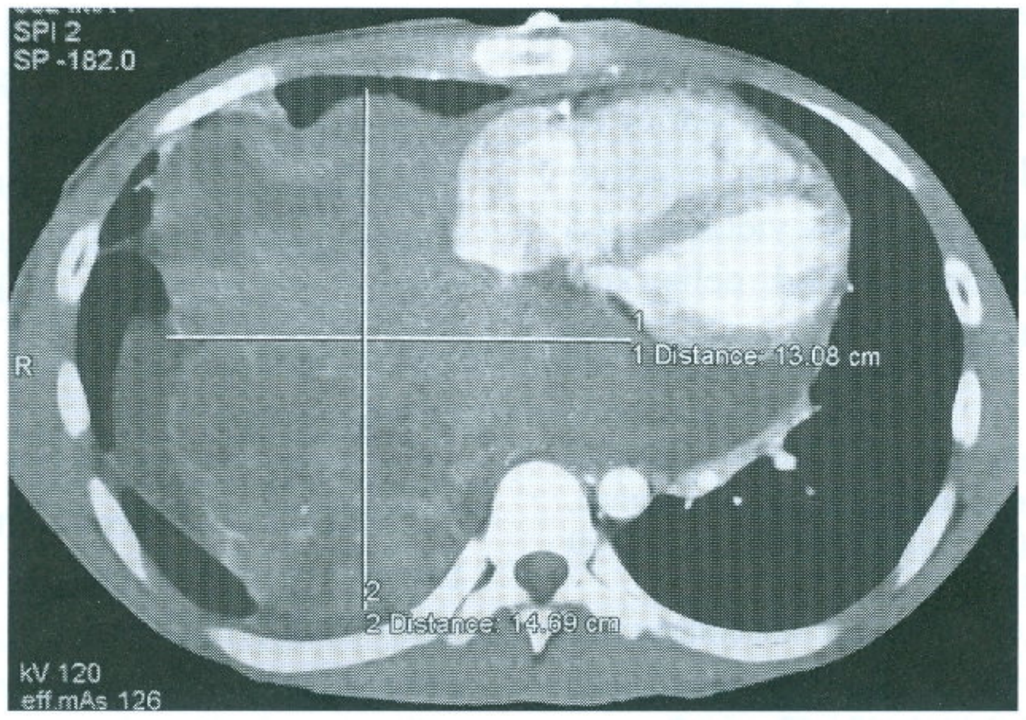
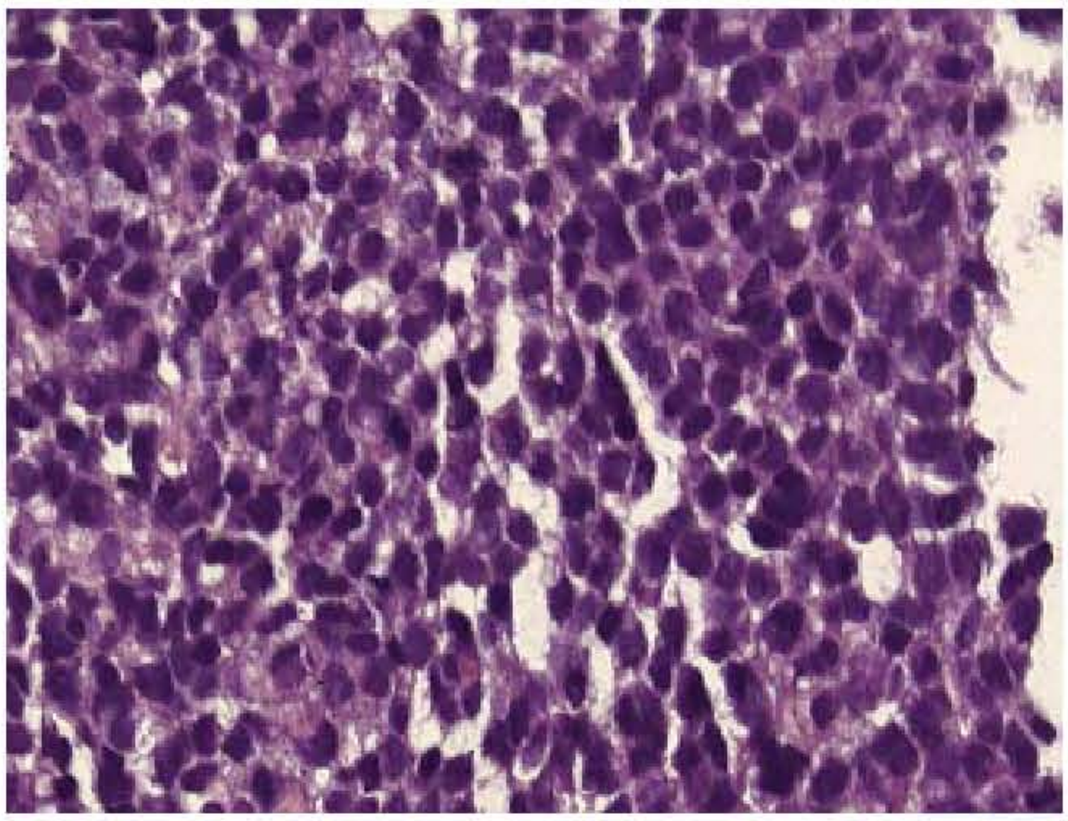


Fig. 2. Large heterogeneous mass in the right lung appearing continuous with the pericardial space and compressing the left atrium.



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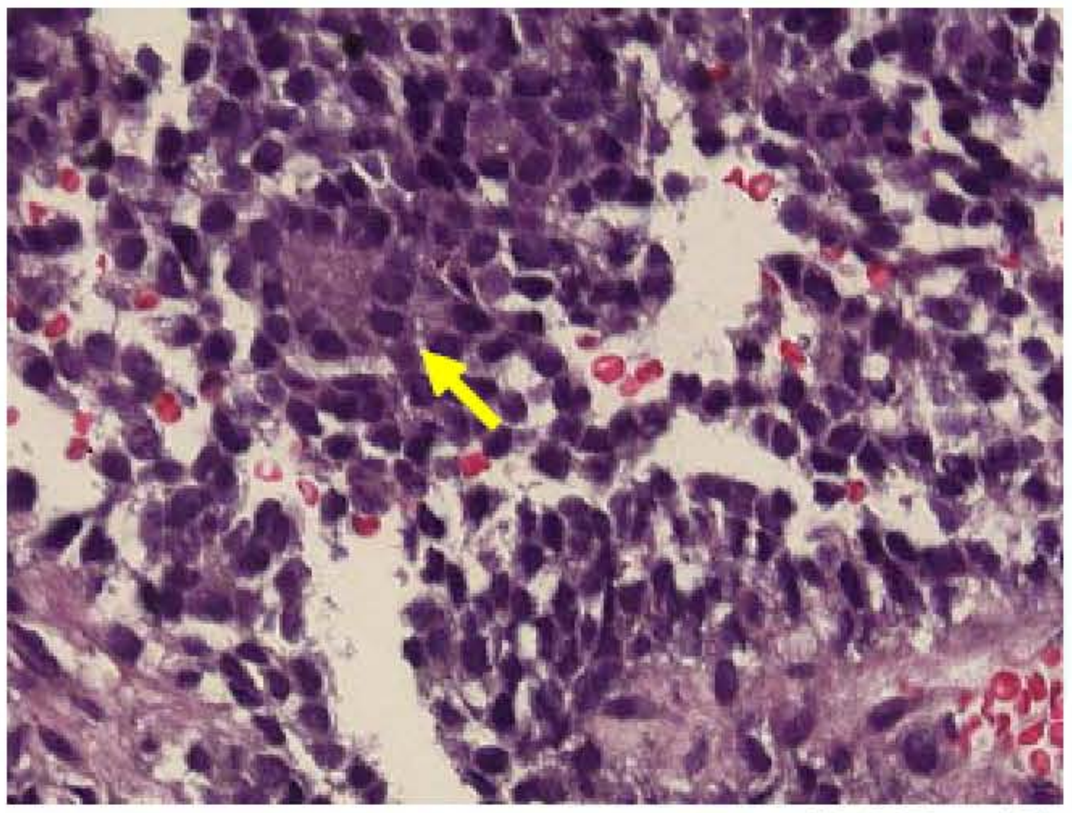

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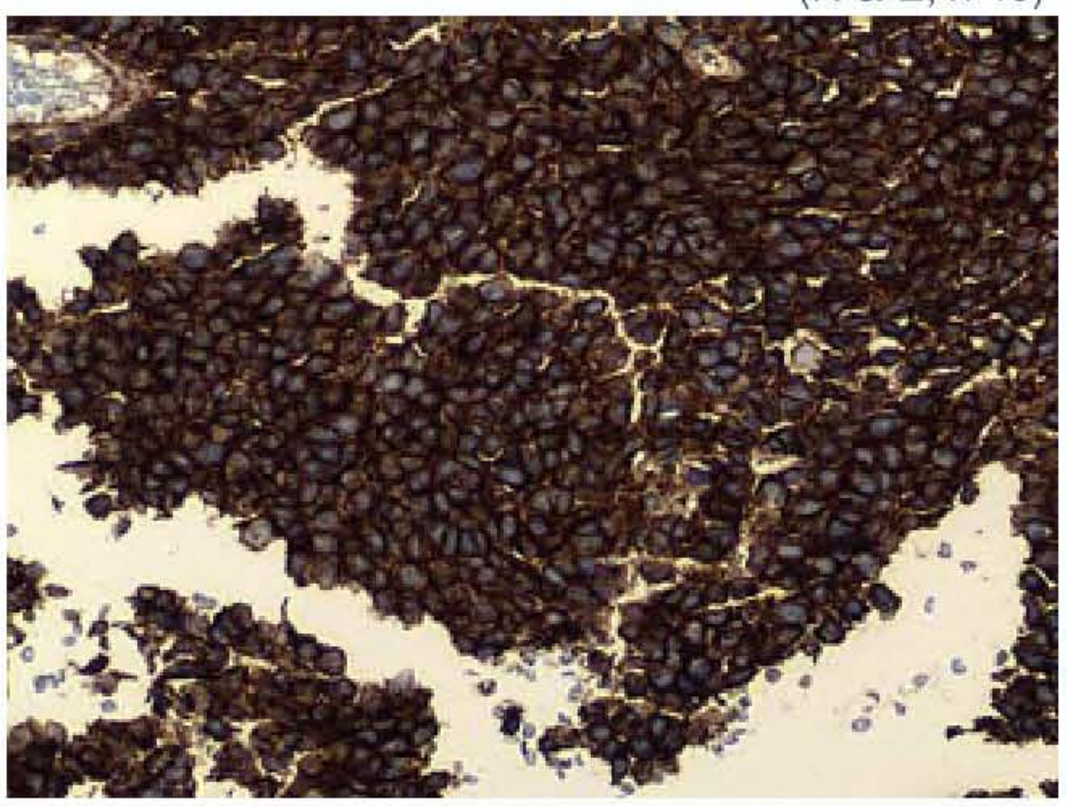
(A) Small blue round cells;
(B) Homer–Wright rosette formation;
(C) CD99 positive staining.



(Dako monoclonal, x 40)

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Fig. 3. Histological slides demonstrating primitive neuroectodermal tumor. (A) Small blue round cells; (B) Homer–Wright rosette formation; (C) CD99 positive staining.