

A Case of Pleural Epithelioid Sarcoma of Proximal Type Presenting as Malignant Pleural Mesothelioma

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CASE REPORT

A 45-year-old man presented with progressing dyspnea and pain in the right shoulder for 1 month. His only medical history was an episode of deep vein thrombosis in the right arm 10 years earlier. He was a smoker with estimated 15 pack-years. He worked as a truck driver and had no history of asbestos exposure. A physical examination by his general practitioner revealed decreased breath sounds over the right lung.

A subsequent chest radiograph revealed a large pleural effusion in the right pleural cavity. On pleuracentesis 3300 ml of bloody pleural effusion was drained. Bacteriology was negative and cytology revealed no malignant cells.

A computed tomography of thorax and abdomen showed several irregular pleural plaques resembling malignant pleural mesothelioma (MPM). No enlarged lymph nodes were seen. A whole-body positron emission tomography-computed tomography showed increased 18F-fluorodeoxyglucose uptake in the pleura of the right hemithorax and in lymph nodes of the mediastinum and the retroperitoneum, as seen in Figure 1. During the subsequent thoracoscopy red and white swollen changes were seen in both parietal and visceral pleura.

Pathology

Pleural biopsies revealed round/polygonal tumor cells in a solid or diffuse pattern with moderate amounts of eosinophilic cytoplasm and central but focally eccentric pleomorphic nuclei (rhabdoid characteristics) with prominent nucleoli and finely granulated chromatin (Fig. 2). Immunostains were strongly positive to mesenchymal vimentin and epithelial pancytokeratin KL-1 and CK8 antigens combined with positive CD34 and loss of INI-1. A focal reaction was noticed to EP4, EMA, CK20, SMA and cytoplasmic WT-1. There was no reaction to the following epithelial markers: carcinoembryonic antigen (CEA), cytokeratin 7 (CK7), thyroid transcription factor-1 (TTF-1), cytokeratin 5 and 6 (CK5/6), tumor protein 63 (P63), homeobox protein CDX-2 (CDX-2), Hep Par-1 (HEP-1), renal cell carcinoma (RCC), cluster of differentiation 10 (CD10), to specific mesothelial markers Calretinin, HBME1, WT-1 (nuclear), D2-40, or to the endothelial markers CD31 and Fli-1, the muscle marker Desmin, the

melanoma markers: S-100, Melan-A, human melanoma black-45 (HMB-45), microphthalmia associated transcription (MITF), the germ cell tumor markers alpha-fetoprotein (AFP), placental alkaline phosphatase (PLAP), human chorionic gonadotropin (HCG), Inhibin, HPL, or the neuroendocrine markers CD56, Chromogranin-A (synaptophysin showed very weak nonspecific focal staining), CD99, CD30, CD117, or Bcl-2. Fluorescent in situ hybridization for deletion of the cyclin-dependent kinase inhibitor 2A (CDKN2A) gene was negative.

This patient showed symptoms only 1 month before admission. His condition rapidly deteriorated and 2 months later he was admitted to a hospice and died within 1 week.

DISCUSSION

These morphological characteristics, immunohistochemistry (IHC), and molecular biology were consistent with epithelioid sarcoma of proximal type, rather than mesothelioma, carcinoma, angiosarcoma, or synovial sarcoma.¹⁻³ A search in PubMed with key words pleural tumor, epithelioid sarcoma, and pleural sarcoma revealed no other cases of primary pleural epithelioid sarcoma.

As a primary in the pleura this is extremely unusual, but perhaps underdiagnosed. Because of its involvement of the whole right pleura and no evidence of a primary tumor anywhere else, we consider it to be a primary in the pleura.

With improved diagnostic tools such as IHC, we might in the future see more cases of pleural epithelioid sarcoma in the pleura, which might in the past have been diagnosed as MPM. In clinical practice this will affect our course of treatment.

MPM and sarcomas do not respond to the same treatment. Cisplatin and pemetrexed are the first-line treatment for MPM, whereas doxorubicin is first-line for treating sarcomas. Second and third lines also differ.

A correct diagnosis is essential for choosing the right treatment and obtaining the best outcome for the patient. With more extensive use of IHC and molecular pathology we can improve our diagnostic precision and thus offer our patients the best treatment.

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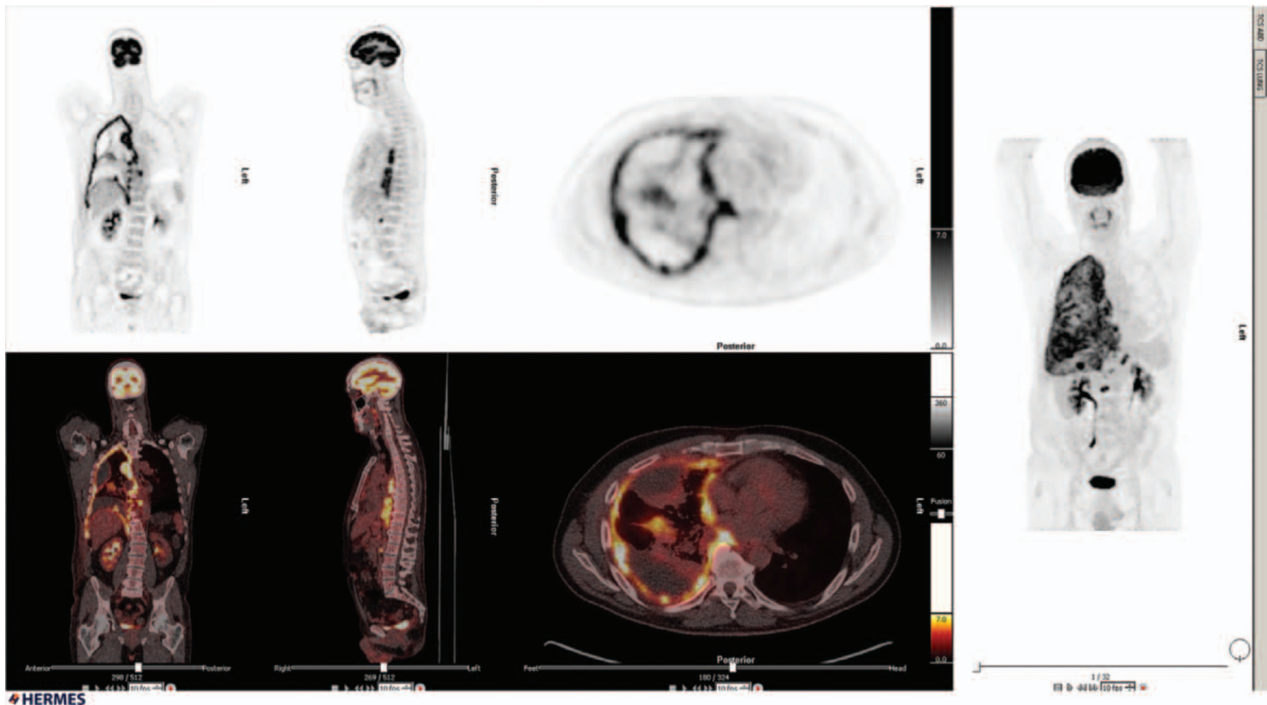


FIGURE 1. Positron emission tomography–computed tomography showing increased ^{18}F -fluorodeoxyglucose uptake in the pleura in the right hemithorax, lymph nodes in the mediastinum and retroperitoneum.

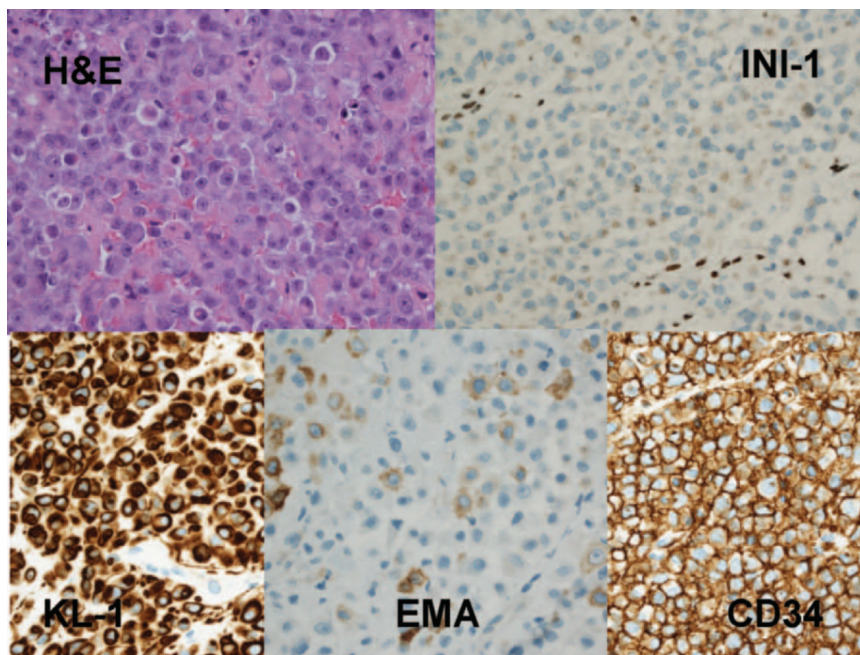


FIGURE 2. Standard hematoxylin and eosin (H&E) and immunostainings for integrase interactor 1 (INI-1), antipancytokeratin (KL-1), epithelial membrane antigen (EMA), and cluster of differentiation (CD34). All at high power ($\times 400$).