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A Rare Case of Sarcomatoid Carcinoma of the Lung with Spine Metastasis, Including a Literature Review

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 63
Final Diagnosis: Sarcomatoid carcinoma of the lung with spine metastasis
Symptoms: Back pain • cough
Medication: —
Clinical Procedure: Lung biopsy • laminectomy
Specialty: Pulmonology

Objective: Rare disease

Background: Sarcomatoid carcinoma is a rare, aggressive, malignant cancer composed of sarcoma and sarcoma-like components, and can occur in different organs such as the thyroid gland, bone, skin, breast, pancreas, liver, urinary tract, and lung. Pulmonary sarcomatoid carcinoma accounts for only a small percentage of lung cancers and has histological variants that include pleomorphic carcinoma, giant cell carcinoma, spindle cell carcinoma, carcinosarcoma, and pulmonary blastoma.





Case Report: Here, we present a case of sarcomatoid carcinoma in a 63-year-old HIV-positive Hispanic male who presented with back pain, dry cough, and weight loss. A CT scan of his chest showed an ovoid mass in the lower lobe of the left lung, and an MRI of the spine showed a left lateral paraspinal soft tissue mass causing central canal stenosis and mild cord compression. The patient underwent laminectomy and resection of the spinal mass. A transthoracic needle biopsy of the lung and spinal masses had similar histopathology, and were indicative of sarcomatoid carcinoma.

Conclusions: We report a rare case of sarcomatoid carcinoma involving both the lung and spinal cord in the same patient. Sarcomatoid carcinomas of the lung have poor prognosis and are aggressive cancers. Moreover, our case also had the co-occurrence of HIV and sarcomatoid carcinoma.

MeSH Keywords: Lung Neoplasms • Neoplasm Metastasis • Sarcoma

Abbreviations: CT – computed tomography; MRI – magnetic resonance imaging

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/904584>

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Background

Sarcomatoid carcinoma is a rare form of cancer in which the cells exhibit properties characteristic of both epithelial and mesenchymal tumors; it can occur in multiple organs, including skin, bone, thyroid, breast, liver, pancreas, urinary tract, and lung [1].

Sarcomatoid carcinoma may consist of a heterogeneous group of non-small cell carcinomas that also contain a component of true sarcoma, or they may be composed in part or completely of sarcoma-like elements. Clinical presentation may vary, including chest pain, dyspnea, cough, and hemoptysis. Sarcomatoid carcinoma is very aggressive, with an overall 5-year survival rate of approximately 20% [2].

Limited data has shown that most cases of sarcomatoid carcinomas occur with advanced local disease and metastasis. With respect to the lung, pulmonary sarcomatoid carcinomas are uncommon, representing less than 1% of all lung cancers [3]. Pulmonary sarcomatoid carcinomas has histological variants that include pleomorphic carcinoma, giant cell carcinoma, spindle cell carcinoma, carcinosarcoma, and pulmonary blastoma [4].

Here, we describe a rare case of a lung mass with spinal metastasis that had histopathologic features of sarcomatoid carcinomas.

Case Report

A 63-year-old Hispanic male presented with back pain, dry cough, and weight loss that had begun 3 weeks prior to his visit. He was not experiencing any of the following symptoms: shortness of breath, chest pain, hemoptysis, weakness of the lower extremities, and urinary or stool incontinence. He was a heavy smoker and his medical history included HIV (CD4 count of 498), hepatitis C, and anal warts.

Upon physical examination, the patient was revealed to be thin, and he appeared alert and comfortable. He was afebrile, his blood pressure was 132/76 mmHg, and his oxygen saturation was 98% on room air. During a respiratory examination, rhonchi were audible, but a spinal exam did not reveal any localized tenderness or swelling. A neurological examination showed no weaknesses or losses of sensations, and no abnormal findings were noted on his cardiac, abdominal, or skin examinations.

Basic laboratory tests, which included a complete blood count, basic metabolic panel, and liver function tests, were all normal. His chest radiograph showed a round consolidation in the

posterior retro cardiac lower left lobe (Figure 1A, 1B). A CT scan of the chest showed heterogeneous soft tissue density, and a mass in the lower left lobe measuring 7.4×6.3 cm in the axial plane and measuring approximately 6 cm from superior to inferior (Figure 2A–2C). Due to his reports of back pain, an MRI was performed of the thoracic spine. The results showed an osseous soft tissue lesion involving the posterior elements of the T8 vertebral body and extending into the posterior epidural space, which resulted in severe central canal stenosis and mild cord compression at this level (Figure 3A, 3B). The neurosurgery team was called, and the patient was started on steroids to alleviate cord compression. A partial laminectomy was performed for T 7 and T 9, and a total laminectomy for T 8, as well as removal of the metastatic tumor, followed by radiation therapy. The histopathologic findings of the tumor were similar to characteristics of lung pathology in sarcomatoid carcinoma.

Subsequently, the patient underwent a PET scan, which showed abnormal hypermetabolic activity in the lower lobe of the left lung and a mass with a standardized uptake value (SUV) of 12.3. In addition, he had a lateral paraspinal soft tissue mass with abnormal hypermetabolic activity and an SUV of 6.1 (Figure 4A, 4B). He later underwent a fiber optic bronchoscopy, with a transbronchial biopsy, which was non-diagnostic. Transthoracic needle aspiration was performed on the mass in the lower left lobe, which revealed tumor cells consisting of malignant spindle cells (sarcoma-like features) within numerous fibrous stroma. The tumor cells were strongly immune-reactive to cytokeratin (CK), CAM 5.2, p40 (focal), and GATA-3, but they were negative for CK7, CK20, thyroid transcription factor 1 (TTF-1), paired box gene 8 (PAX8), caudal type homeobox 2 (CDX2), CD34, and desmin. The morphology and staining pattern were consistent with a sarcomatoid carcinoma subtype pleomorphic carcinoma (Figure 5A–5C).

Due to the extensive nature of the tumor, the patient was deemed unsuitable for resection of the lung mass. Instead, he was started on chemotherapy consisting of Gemcitabine/Carboplatin, to followed by continued maintenance with Gemcitabine. In addition, he was started on bisphosphonates for bone metastasis. Currently, he receives treatment from the pulmonary and oncology clinics as an outpatient.

Discussion

This case report describes the rare occurrence of lung mass with spinal metastasis, with histopathologic findings similar to those of sarcomatoid carcinoma. Our report also shows a co-occurrence of HIV and sarcomatoid carcinoma. Therefore, this case raises the possibility that sarcomatoid carcinoma and HIV are associated, although more studies are needed to confirm this.

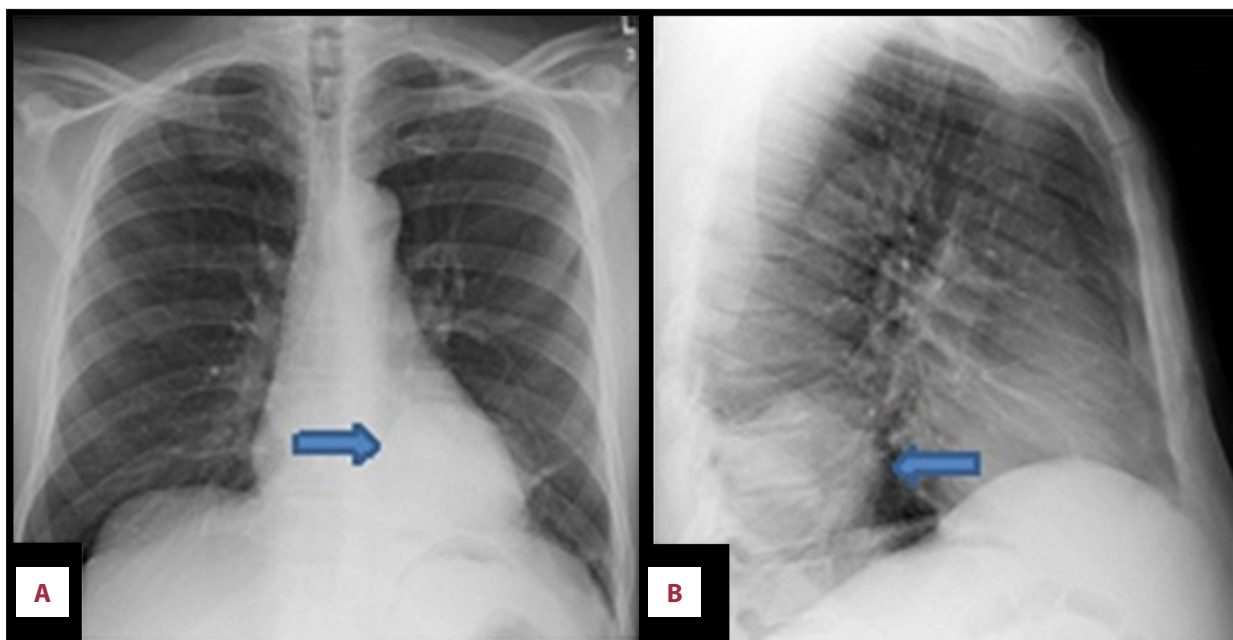


Figure 1. Chest radiographs of the patient. (A) PA view and (B) lateral view of the patient's chest showing a retro-cardiac ovoid mass (arrows) in the lower left lobe.

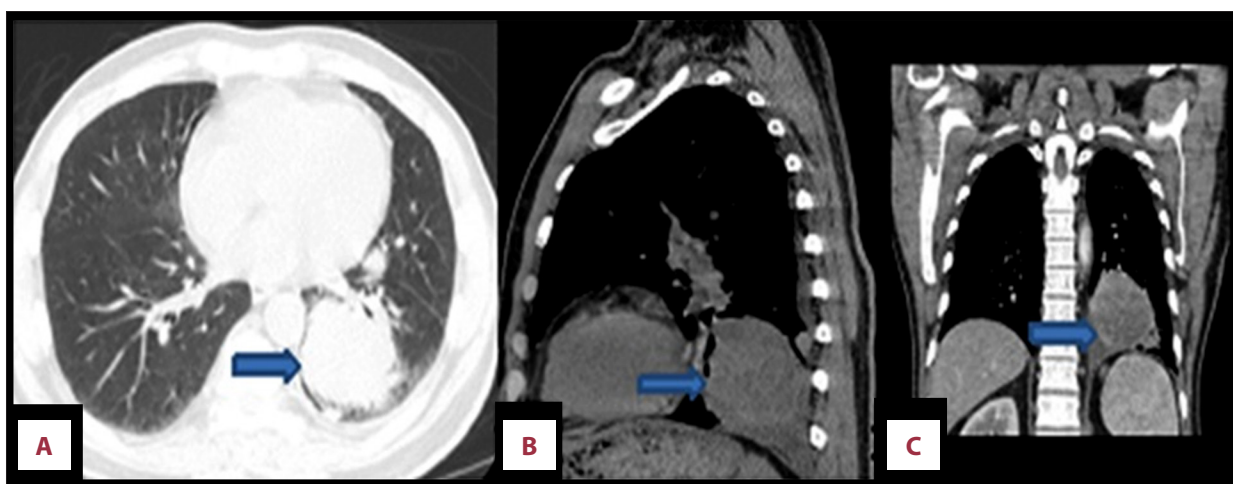


Figure 2. CT scans of the chest. The (A) axial, (B) sagittal, and (C) coronal views showing an ovoid mass (arrows) in the lower left lobe measuring 7.4x6.3 cm in the axial plane by approximately 6 cm from superior to inferior.

Sarcomatoid carcinomas are unique among lung carcinomas in that, although they are considered carcinomas, they contain cytological and tissue architectural features that are usually characteristic of sarcoma [5,6]. In 1981, sarcomatoid carcinomas that corresponded to spindle cell were categorized as a variant of squamous cell carcinoma [7]. In 1999, carcinomas with spindle and/or giant cells were classified under the heading of “carcinomas with pleomorphic, sarcomatoid, or sarcomatoid elements” [8]. Moreover, the World Health Organization 2004 classification defined pulmonary sarcomatoid carcinomas as “poorly differentiated non-small cell carcinoma that have a histological appearance suggesting mesenchymal differentiation” [9].

As of 2015, the World Health Organization had no changes to diagnostic criteria nor terminology for these tumors since the 2004 Classification, but recommends molecular testing to the known genetic abnormalities with histologic findings. It is difficult to diagnose these tumors using small biopsy samples [10].

Sarcomatoid carcinoma is a subtype of non-small cell lung cancer, and is defined by the presence of a sarcoma or sarcoma-like component, and includes 5 subtypes: (a) spindle cell carcinoma, (b) giant cell carcinoma (a tumor almost entirely composed of giant cells), (c) pleomorphic carcinoma, (d) carcinosarcoma (a mixture of non-small cell lung cancer and sarcoma containing

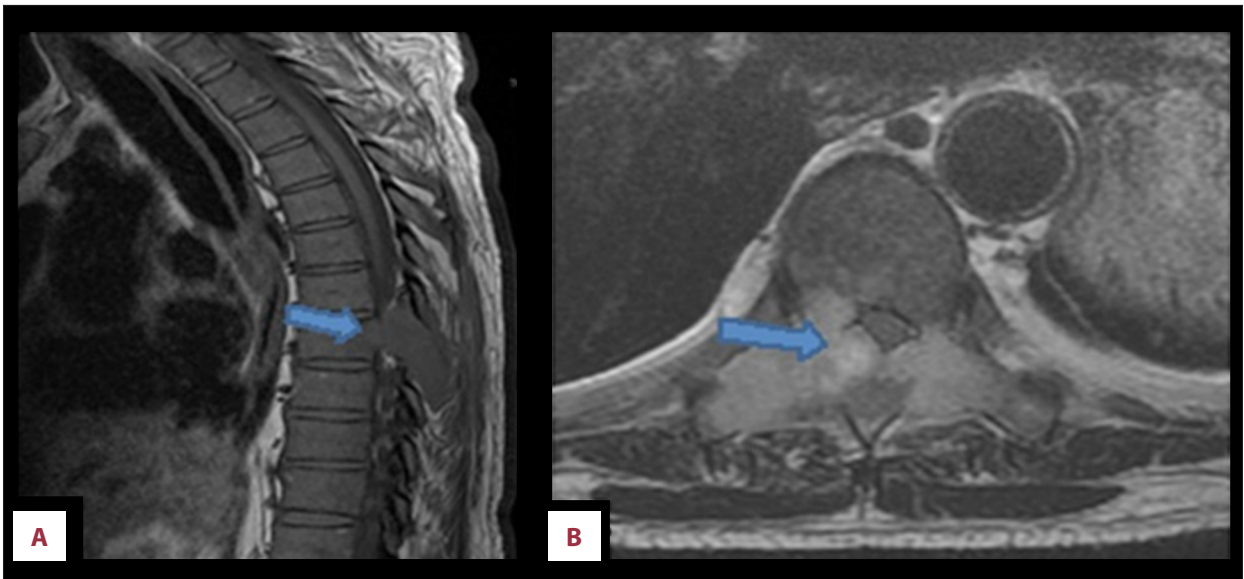


Figure 3. MRI images of the thoracic spine. Images in (A) and (B) show an osseous soft tissue lesion (arrows). The lesion involved the posterior elements of the T8 vertebral body, extending into the posterior epidural space, and resulted in severe central canal stenosis and mild cord compression at this level.

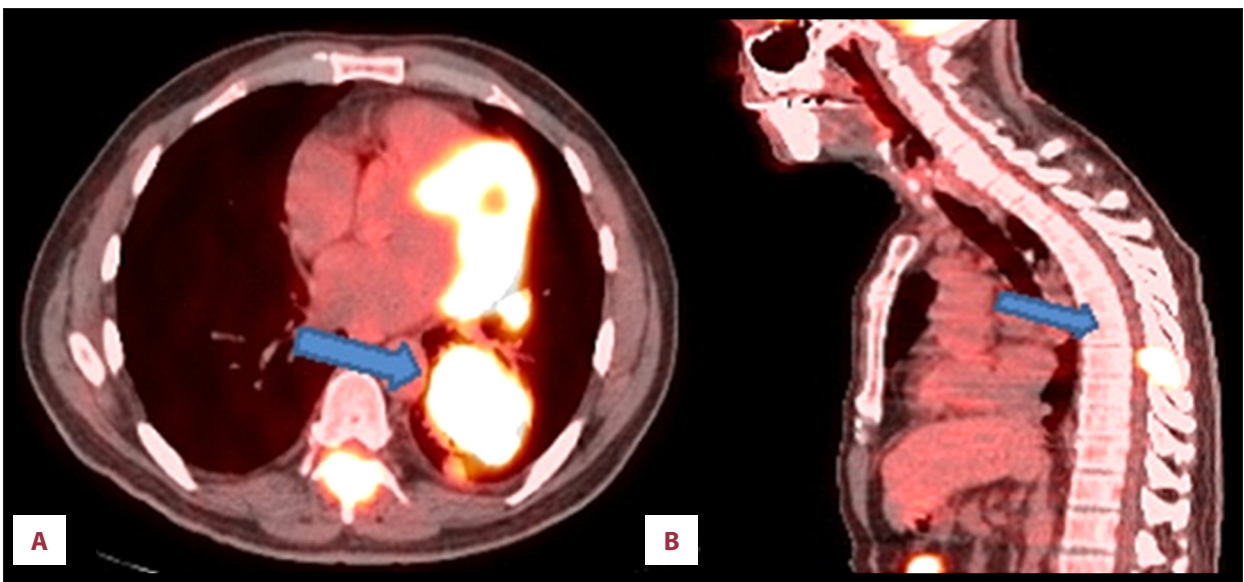


Figure 4. PET scans. (A) Abnormal hypermetabolic activity was identified in a mass (arrow) in the lower lobe of the left lung. (B) A paraspinal soft tissue mass (arrow) with abnormal hypermetabolic activity was identified.

heterologous elements), and (e) biphasic pulmonary blastoma (a tumor composed of embryonal-type epithelial elements and primitive mesenchymal stroma). Sarcomatoid carcinoma is more common in men who are smokers, and the average age of individuals at diagnosis is 65 years, except for the pulmonary blastoma subtype, for which the average age at diagnosis is 35 years. Multiple risk factors have been associated with sarcomatoid carcinoma of the lung, such as smoking cigarettes, cigars, or pipes, and exposure to asbestos in building construction and electrical insulation [11].

There is no specific clinical presentation, although patients may present with cough, dyspnea, hemoptysis, chest pain, or weight loss [12]. Sarcomatoid carcinoma is characterized by rapid growth, invasion, disease recurrence, and metastases. Pulmonary sarcomatoid carcinoma presents as either a peripheral or central lesion and grows by invading the bronchial tree, the pulmonary parenchyma, and the adjacent anatomical structures (mediastinum and chest wall) in the form of widely necrotic and hemorrhagic, round-to-bosselated large masses [13].

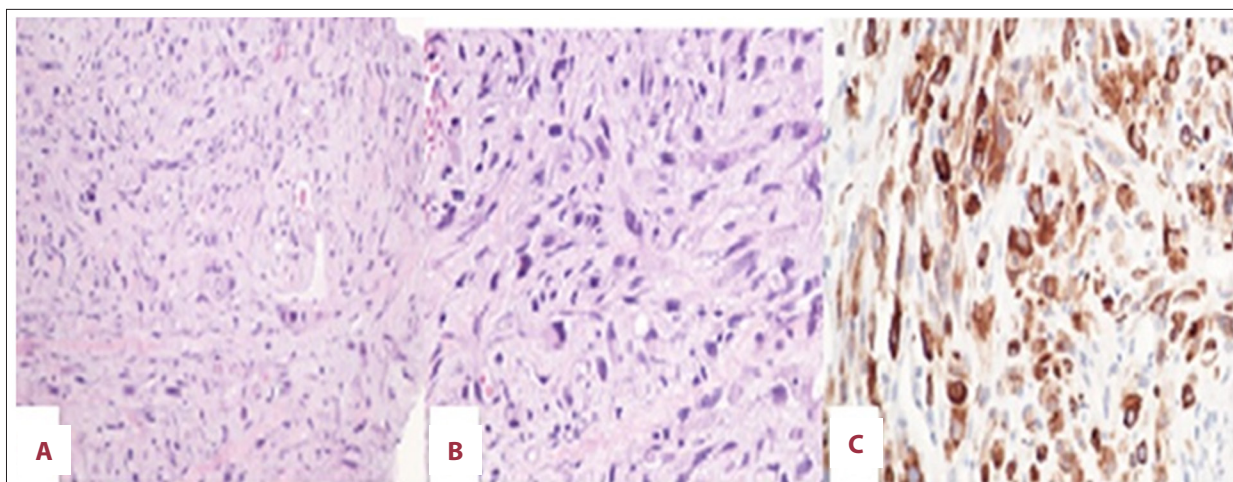


Figure 5. Results of immunohistochemical staining. (A) The tumor cells consisted of malignant spindle cells (sarcoma-like features) within abundant fibrous stroma (HE stain; magnification $\times 200$). (B) The malignant spindle cells showed nuclear polymorphism and mitotic figures (HE stain; high magnification $\times 400$). (C) The tumor cells were strongly immunoreactive to cytokeratin (CAM 5.2) stain.

While light microscopy is sufficient to diagnose most of these tumors, immunohistochemistry can be useful in selected settings [14]. Immunohistochemical investigations can be used to help determine the histological type of non-small lung carcinoma. In general, CK5/6 and p63 are markers of squamous cell carcinoma, whereas SP-A and TTF-1 are markers of adenocarcinoma. Staining with CK7 and CK20 antibodies can help discriminate between primary lung carcinoma and metastatic lung carcinoma. In addition, pan cytokeratin (CAM 5.2 and LP 34) has been reported to be present in sarcomatoid carcinoma of the lung [15].

A study by Kim et al. showed Napsin-A (81%) and TTF-1 (70%) was positive in adenocarcinoma, but only 2% had TTF-1 positive squamous cell carcinoma. Immunohistochemical stain p63 (91%) and CK 5/6 (90%) are sometimes (9%) positive in squamous cell carcinoma and are rarely (4%) positive in adenocarcinoma ($P < .001$) [16]. Brandler et al. showed that GATA 3 and P40 immunohistochemical stain was positive in urothelial adenocarcinoma [17]. P40 is a more sensitive and specific immunohistochemical marker for pulmonary squamous cell carcinoma over p63 [18].

There are no pulmonary sarcomatoid carcinoma prospective studies to date, primarily due to the low incidence of this cancer, as well as the difficulties in diagnosing it. Since the cancer is so aggressive, most patients are diagnosed very late, in the advanced stages. There are only a few retrospective studies that compared sarcomatoid carcinoma to conventional non-small cell cancer and, reporting that patients with sarcomatoid carcinoma did significantly worse in terms of median survival, disease progression, and overall survival [19].

In a study by Ro et al., a tumor size of > 5 cm, a clinical stage of > 1 , metastasis, associated genetic mutation (K-Ras or p53 mutation), and lymph node involvement significantly shortened patient survival. Moreover, sarcomatoid carcinoma had a median survival of 10 months, which was much shorter than for other lung carcinomas (20 months for adenocarcinoma, 12.6 months for large cell carcinoma, and 18.5 months for squamous cell carcinoma) [20].

In cases where the tumor is localized, surgery is an adequate course for treatment. In contrast, since no data are currently available for the metastatic disease, patients are treated as having non-small cell lung cancer [21]. A study by Vieira et al. showed that the progression-free survival was not statistically significantly different between patients who received platinum-based chemotherapy versus those who did not. Additionally, no statistically significant difference in overall survival was observed (7 months with platinum versus 5.3 months without; $P = 0.096$) [22].

Our patient was a male smoker, which is risk factor for developing sarcomatoid cancer. Also, immune histochemical stains for lung mass and spinal mass were positive for cytokeratin and CAM 5.2, which favor sarcomatoid cancer of the lung. As the disease was not localized, our patient was not a candidate for surgical resection of the lung mass. Other lung cancers like adenocarcinoma, squamous cell, large cell, and small cell have been reported in HIV patients, but our case shows there could be an association between HIV and sarcomatoid cancer.

Conclusions

Sarcomatoid carcinomas of the lung are aggressive cancers with a poor prognosis. The histological and immunohistochemical characteristics are very different from those of non-small

cell lung carcinoma. Our case is unique in that pulmonary sarcomatoid carcinoma is a rare variety of lung carcinoma with spinal metastasis, and our case also had co-occurrence of HIV and sarcomatoid carcinoma.

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