Case Report

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Case Report: The First Documented Case of Advanced Metastatic Dermatofibrosarcoma Protuberans (DFSP) with Extensive Intrathoracic Dissemination and Mediastinal Shift in West Africa

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive cutaneous malignancy with a low propensity for distant metastasis. When systemic dissemination occurs, it primarily targets the lungs, often secondary to a fibrosarcomatous transformation (FS-DFSP). We report the case of an 81-year-old Gambian male presenting with severe progressive dyspnea. He had a 14-year history of recurrent, disseminated cutaneous tumors across his limbs and torso, histologically confirmed as DFSP. Advanced thoracic imaging revealed a massive, space-occupying left intrathoracic mass causing complete lung collapse and a severe rightward mediastinal shift, alongside contralateral pulmonary nodules. To our knowledge, this represents the first documented report of advanced metastatic DFSP with extensive intrathoracic dissemination in West Africa. This case highlights the profound diagnostic challenges associated with the slow-growing nature of the tumor and the clinical difficulty less experienced physicians face in linking indolent dermatological conditions to internal organ affection. Furthermore, it underscores the logistical hurdles of managing complex soft-tissue sarcomas in resource-limited environments where specialized diagnostic validation and targeted therapies are unavailable.

Keywords: Dermatofibrosarcoma protuberans, rare diseases, cancer, west Africa and Gambia.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon dermal soft-tissue sarcoma, representing approximately 0.1% of all malignancies and about 1% of soft-tissue sarcomas, with a global incidence estimated between 1 and 5 cases per million persons-years [1]. Notable demographic disparities exist, with incidence reported to be "1.14 times higher in women than in men, and nearly two times higher in Black individuals compared to White individuals," while long-term outcomes remain excellent, reflected in a "10-year relative survival rate of 99.1%." Despite this favorable prognosis, specific factors—including advanced age, male sex, Black race, and tumors arising on the limbs or head rather than the trunk—are associated with increased all-cause mortality. DFSP is classically indolent and slow-growing but highly infiltrative locally, with a strong propensity for recurrence after inadequate excision, whereas distant metastasis is historically rare, occurring in fewer than 5% of cases; when it does occur, "the lungs represent the undisputed primary site of involvement," particularly in the context of fibrosarcomatous transformation (FS-DFSP), the aggressive histologic variant most strongly linked to metastatic spread.

Although DFSP is well documented globally, published accounts of advanced thoracic dissemination remain virtually non-existent in West African medical literature [2], underscoring the rarity and clinical significance of such presentations in this region.

Case Presentation

Patient History and Clinical Presentation

An 81-year-old Gambian male presented to the medical clinic with a chief complaint of progressive shortness of breath and difficulty breathing. The dyspnea developed gradually over several months but experienced a severe, debilitating exacerbation over the preceding 3 weeks. The patient's past medical history was significant for multi-focal, disseminated cutaneous tumors distributed across his chest, back, and extremities. The primary lesion emerged approximately 14 years prior and was surgically excised due to its obstructive size; however, no histopathological feedback or formal diagnosis was provided to the patient at that time. Over the subsequent years, multiple new cutaneous nodules emerged, exhibiting an extremely slow, indolent growth pattern. A diagnostic biopsy performed just one year ago and formally established a diagnosis of dermatofibrosarcoma protuberans.

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Figure 1: Posterior view

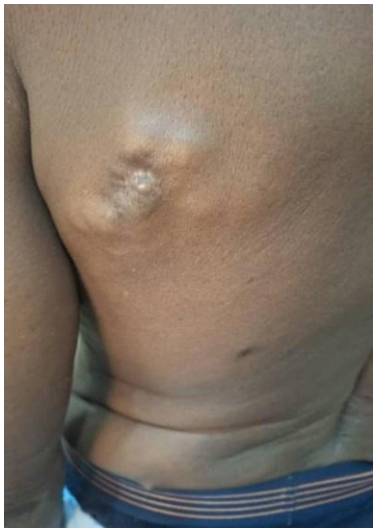


Figure 2: Anterior view

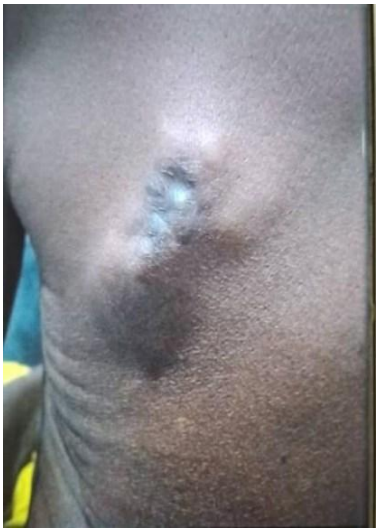
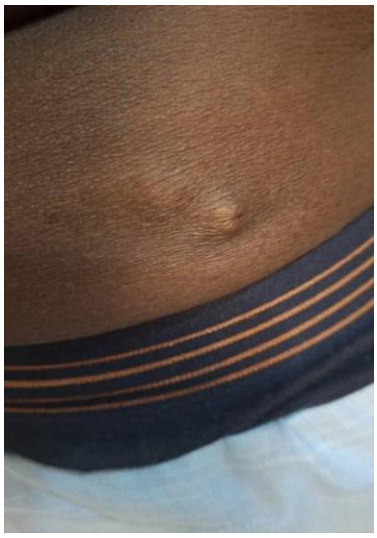


Figure 3: Anterior lower view



Chest Assessment: Inspection revealed globally reduced chest expansion, which was profoundly marked on the left hemithorax. Palpation exposed a marked increase in tactile vocal fremitus ipsilateral. Upon auscultation, vesicular breath sounds were completely absent throughout the left lung field, accompanied by scattered, low-pitched expiratory wheezes in right lung.

Diagnostic Imaging

Advanced thoracic imaging, including Computed Tomography (CT), delineated a catastrophic degree of intrathoracic tumor progression: Figure 4-5.

- **Left Hemithorax:** A massive, homogeneous, space-occupying intrathoracic soft-tissue mass filled the entirety of the left thoracic cavity, resulting in atelectasis of the left lung parenchyma.
- **Mediastinal Shift:** The extensive tumor volume exerted a severe mass effect, causing a massive rightward shifting of vital mediastinal structures, including the heart, trachea, and great vessels.
- **Contralateral Lung:** Advanced dissemination confirmed by the presence of well-defined, metastatic pulmonary nodules within the partially compressed right lung fields.

Figure 4: Chest CT scan coronal view

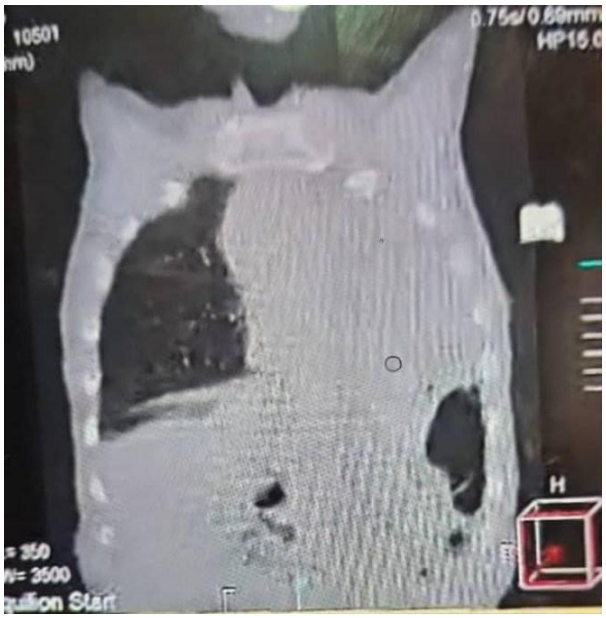
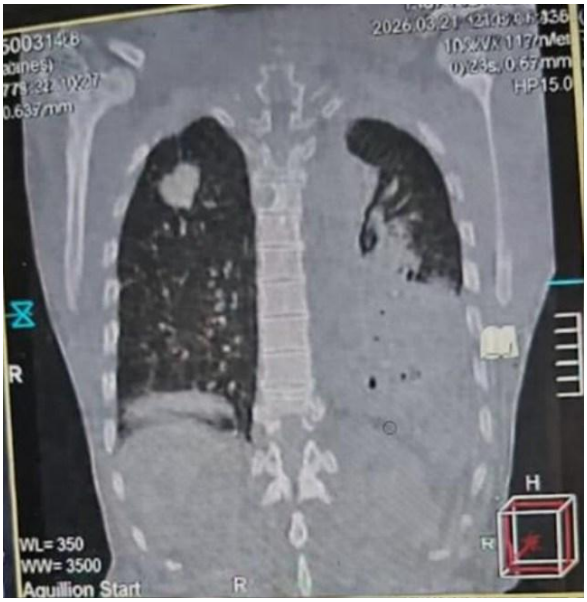


Figure 5: Chest CT scan coronal view



Histopathological Report: Department of Pathology and Laboratory Medicine at the EFSTH. Hosp No: 11345084
Lab No FNAB: 24.12.375
Fine Needle Aspiration Cytology (FNAC) of the subcutaneous nodules showed cellular smears composed of atypical spindle-to-polygonal mesenchymal cells with hyperchromatic nuclei and scanty cytoplasm. The pathological features were interpreted as an intermediate-grade mesenchymal neoplasm consistent with a recurrence of DFSP. Intermediate grade mesenchymal neoplasm.

Discussion

A primary diagnostic challenge illustrated by this case is the incredibly slow, indolent growth rate characteristic of early-stage DFSP. Because these tumors typically develop over many years as painless, superficial nodules or plaques, they are frequently dismissed by both patients and clinicians as benign dermatological entities like lipomas, keloids, or dermatofibromas. Patients often ignore these early lesions for over a decade, allowing the tumor to silently infiltrate underlying tissues or slowly progress systemically before seeking formal medical attention. This delayed clinical presentation contributes significantly to advanced tumor burden upon initial comprehensive staging. This case underscores an essential educational gap in general clinical practice: The difficulty less experienced physicians face in connecting seemingly localized dermatological conditions with severe, internal visceral organ affection [3]. In standard clinical workflows, superficial skin lesions are managed independently of systemic symptoms. Without specialized oncological experience, it is highly challenging for a primary care clinician to realize that a patient's worsening dyspnea is directly tied to a slow-growing skin mass diagnosed months prior. This diagnostic disconnect often leads to fragmented care, where the respiratory symptoms are initially mismanaged as primary pulmonary diseases (such as tuberculosis, bacterial pneumonia, or congestive heart failure), delaying the vital thoracic imaging needed to reveal the underlying sarcoma metastasis. As the first documented report of this advanced manifestation in West Africa, this case highlights a critical health equity gap. Confirming the COL1A1-PDGFB fusion gene via FISH or RT-PCR is required to predict targeted treatment success [3,4], yet neither molecular diagnostics nor targeted TKI therapies like Imatinib are accessible or affordable in The Gambia. Managing such complex, systemic disease demands a multidisciplinary tertiary infrastructure [5,6] that remains largely unavailable in the sub-region.

Conclusion

This represents the first reported case of advanced metastatic DFSP presenting with massive intrathoracic extension and structural mediastinal shift in West Africa. The case provides a stark lesson on the dangers of diagnostic oversight stemming from the tumor's deceptive, slow growth and the clinical difficulty of linking skin lesions to internal organ pathology. Addressing these diagnostic gaps and mitigating therapeutic resource disparities are essential to improving outcomes for rare soft-tissue malignancies in resource-constrained settings.

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