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Pleomorphic Spindle Cell Rhabdomyosarcoma of Retroperitoneum in Older Adult: A Rare Case Entity

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KEYWORDS

Spindle cell rhabdomyosarcoma, retroperitoneum, surgical resection

ABSTRACT:

Background:

Rhabdomyosarcoma (RMS) is a rare and aggressive malignant neoplasm derived from primitive mesenchymal cells capable of skeletal muscle differentiation. Although it predominantly occurs in children and young adults, spindle cell RMS, a rare histological variant, has been described in adults with distinct clinical and pathological features. Retroperitoneal RMS is particularly uncommon and often presents at an advanced stage due to the deep-seated location, making early detection and complete surgical excision difficult.

Case Presentation:

We describe the case of a 70-year-old male who presented with progressive abdominal discomfort and fullness. Computed tomography (CT) of the abdomen revealed a large retroperitoneal mass displacing adjacent structures and encasing major vascular channels. The patient underwent exploratory laparotomy, during which incomplete resection was performed because of the tumor's intimate involvement with vital vessels. Grossly, the tumor displayed areas of necrosis and calcification. Microscopic examination revealed spindle-shaped tumor cells arranged in fascicles with evidence of myxoid degeneration, calcification, and extensive necrosis. Immunohistochemistry confirmed the diagnosis of spindle cell RMS, with tumor cells showing positivity for muscle-specific actin, desmin, and vimentin. At the time of surgery, there was no evidence of distant or nodal metastasis.

Conclusion:

This case underscores the diagnostic and therapeutic challenges of retroperitoneal spindle cell RMS in elderly patients. The rarity of this presentation, coupled with the tumor's aggressive nature and tendency to involve major vessels, limits the feasibility of complete resection and complicates management. Early detection through vigilant imaging and timely intervention, along with a multidisciplinary treatment strategy incorporating surgery, chemotherapy, and radiotherapy, remains essential for improving prognosis in such rare cases.

Introduction

Rhabdomyosarcoma (RMS) is a soft tissue malignancy that arises from the primitive rhabdomyoblast of mesenchymal origin. According to the WHO Classification 2013, the RMS is classified as alveolar, embryonal, pleomorphic, and spindle cell RMS with varying differentiation, aggressiveness, and prognosis [1]. Spindle cell RMS is a rare histological variant of RMS. In the early period, it was included as a subtype of embryonal RMS, and now it has been recognized as a distinct subtype of RMS [2]. The incidence rate of spindle RMS accounts for 5-10% with a male preponderance [3]. The most common site of the spindle cell RMS is the head and neck, and other locations such as the diaphragm, uterus, and para testicular regions are also reported in the literature [4]. The spindle cell RMS at the retroperito¬neum is a rare anatomical site, and only a few cases are reported in the literature [5,6]. These rare site lesions in adults show distinct clinical-pathological features with aggressive

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clinical forms compared to other forms in adults and pediatric cases [2]. We report a case of this rare tumor presenting in a 70-year-old male as a progressively increasing abdominal distension with discomfort and posed a challenge at the time of diagnosis.

Case Report

We describe a rare case of spindle cell RMS of the retroperitoneum in a 70-year-old man who presented with complaints of abdominal distension, abdominal discomfort, and a history of decreased appetite for 6 months. Later onset of breathlessness, backache & right thigh numbness. Computed tomography showed a huge tumor mass, measured ~ 22 x 22 x 26 cm (AP x Trans X CC) in size, arising in retroperitoneal space (Figure 1). The patient underwent an Exploratory Laparotomy with the excision of a tumor (Figure 2). The tumor was incompletely excised because of the encasement of significant vessels at the lower pole (Figure 3).

Histologically, the tumor consisted of spindle-shaped (resembling rhabdomyoblast and strap cells) Cells with epithelioid nuclei, prominent nucleoli (Figure 4), Bizarre cells, Binucleated and multinucleated tumor cells (Figure 5), extensive areas of hyalinization, myxoid degeneration, (Figure 6) and calcification was evident.

Immunohistochemically, tumor cells were stained diffusely positive for muscle-specific actin, desmin, and vimentin, scattered positive for myogenin, MyoD1, and myoglobin, with a Ki-67 (MIB-1) proliferative labeling index of 30-35%. Other markers, such as S100 protein smooth muscle-specific actin, were negative in the tumor cells.

Discussion

Rhabdomyosarcoma (RMS) is a highly aggressive and malignant soft tissue tumor that originates from primitive mesenchymal cells, which have the potential to differentiate into skeletal muscle cells. Although RMS is typically associated with childhood, this malignancy in adults, especially the spindle cell variant, remains rare and presents unique clinical challenges [5]. The case of a 70-year-old male with a spindle cell RMS arising from the retroperitoneum provides an opportunity to discuss this rare variant of RMS, its clinical features, diagnostic challenges, management strategies, and prognostic factors.

Regarding anatomical sites, RMS typically occurs in the head and neck, genitourinary tract, and extremities [7]. However, when spindle cell RMS arises in the retroperitoneum, it presents an unusual and rare challenge. Retroperitoneal RMS is less common than other soft tissue sarcomas, often leading to a delayed diagnosis due to its nonspecific clinical presentation. In the reported case, the 70-year-old male patient presented with progressively increasing abdominal distension, abdominal discomfort, and decreased appetite over six months. Other symptoms included breathlessness, backache, and right thigh numbness, complicating the clinical picture [8].

The clinical presentation of spindle cell RMS in the retroperitoneum can often mimic other abdominal masses or gastrointestinal conditions, resulting in a delay in diagnosis. In the present case, the patients had abdominal distension and discomfort, which are common complaints in a wide variety of abdominal pathologies, including benign conditions like gastrointestinal bloating, infections, or even malignancies such as gastrointestinal stromal tumors. Due to this, the clinical symptoms may be nonspecific and do not immediately point toward a diagnosis of RMS, particularly in older adults.

The physical examination may reveal a palpable mass, but this may not be possible in deep-seated lesions like retroperitoneal RMS. Imaging modalities such as computed tomography (CT) or magnetic resonance imaging are crucial in diagnosing retroperitoneal tumors. In the present case, CT imaging revealed a large tumor mass measuring approximately 22 x 22 x 26 cm, which confirmed the presence of a retroperitoneal lesion. Earlier reports also confirmed that CT is an effective imaging modality in diagnosing retroperitoneal RMS [9].

Surgical resection is the mainstay for the spindle cell RMS. However, achieving the complete challenge since it is closely associated with vital structures such as blood vessels or organs. In the present case, the tumor was incompletely excised due to the encasement of significant vessels at the lower pole, a common challenge when dealing with retroperitoneal tumors.

Incomplete resection often increases the risk of recurrence, and the decision to leave behind portions of the tumor must be carefully weighed against the risk of

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postoperative complications, including bleeding, organ damage, or loss of function.

The fact that this patient's tumor was incompletely excised due to its location underscores the aggressive nature of retroperitoneal RMS. Surgical approaches to retroperitoneal tumors must involve careful preoperative planning, and multidisciplinary teams, including surgeons, oncologists, and radiologists, are essential for optimal management.

Histologically, spindle cell RMS consists of spindle-shaped cells that resemble rhabdomyoblasts. These cells have distinct characteristics, such as epithelioid nuclei, prominent nucleoli, and bizarre multinucleated or binucleated cells. The tumor also exhibits hyalinization, myxoid degeneration, and necrosis, all commonly observed in spindle cell RMS, providing valuable diagnostic clues [10]. The extensive areas of calcification and tumor necrosis seen in the current case further support the diagnosis of RMS, as these features are often associated with aggressive sarcomas.

Immunohistochemistry is crucial in confirming the diagnosis and differentiating RMS from other soft tissue tumors. In this case, the tumor cells were positive for muscle-specific actin, desmin, and vimentin, all markers typically expressed in RMS. Though scattered, the presence of myogenin, MyoD1, and myoglobin further supports the diagnosis of RMS, as these are markers indicative of muscle differentiation [11]. The Ki-67 (MIB-1) proliferative index of 30-35% indicates a moderately high rate of cellular proliferation, consistent with the aggressive nature of spindle cell RMS [6].

The tumor's negative staining for S100 protein and smooth muscle-specific actin helps exclude other soft tissue neoplasms, such as malignant peripheral nerve sheath tumors and leiomyosarcomas, which can share similar histological features with RMS. Immunohistochemistry thus provides essential information that aids in confirming the diagnosis and ruling out other potential diagnoses.

The prognosis for patients with spindle cell RMS largely depends on factors such as tumor size, location, extent of surgical resection, and the presence of metastatic disease. Spindle cell RMS in adults tends to have a poorer prognosis compared to other subtypes of RMS, primarily due to the tendency of these tumors to present at later stages and the difficulty in achieving complete resection. Additionally, the retroperitoneum, as a deep anatomical site, complicates diagnosis and treatment, further contributing to the tumor's aggressive clinical course.

The prognosis for this patient is influenced by the incomplete resection, as well as the large size of the tumor at the time of diagnosis. The lack of apparent metastatic disease at the time of surgery is favorable, but close follow-up is essential to monitor for recurrence. Adjuvant therapies, such as radiation therapy or chemotherapy, may be considered in cases where complete surgical resection is not possible. However, their effectiveness in adult patients with retroperitoneal RMS remains controversial.

4. Conclusion

Spindle cell RMS is a rare and aggressive variant of RMS that presents unique diagnostic and therapeutic challenges, primarily when located in the retroperitoneum. This case report highlights the importance of a high index of suspicion for rare malignancies, such as spindle cell RMS, in adult patients with nonspecific abdominal symptoms. Early diagnosis through imaging and histopathological examination is crucial for guiding treatment decisions, and surgical resection remains the cornerstone of management. Although the prognosis for spindle cell RMS in adults is generally poor, the case also emphasizes the need for ongoing research to understand this rare tumor's biology better and identify optimal therapeutic strategies.

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Journal of Chemical Health Risks

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