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Differential Diagnosis of a Pediatric Ovarian Tumor: The Pitfall of Juvenile Fibrosarcoma Versus Spindle Cell Embryonal Rhabdomyosarcoma

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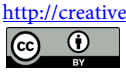
Abstract

Introduction: Ovarian tumors in children are rare, with a malignant rate of 10% - 20%. Beyond common epithelial or germ cell tumors, mesenchymal tumors like spindle cell embryonal rhabdomyosarcoma (ERMS) present significant diagnostic challenges due to their histological similarity to juvenile fibrosarcoma. This study reports a case of a 7-year-old girl to highlight how morphological overlap can lead to diagnostic errors and the critical necessity of immunohistochemistry (IHC) in establishing a diagnosis. **Methods:** A study was performed on a 7-year-old patient who was initially diagnosed with juvenile fibrosarcoma. Following the patient's death, a morphological review was conducted to challenge the initial findings. The diagnostic process involved expanding the immunohistochemical panel from just CD34 and S100 to include myogenin, MyoD1, desmin, EMA, cytokeratin AE1/3, and vimentin to explore the possibility of ERMS. **Results: Clinical Presentation:** The patient presented with abdominal pain, dysuria, and a 9 cm right ovarian mass invading the peritoneum.



Initial Findings: A diagnosis of juvenile fibrosarcoma was suggested based on morphology, and the patient received three cycles of Doxorubicin and Cisplatin. **Outcome:** The patient died two weeks after the third chemotherapy cycle. **Morphological Review:** Re-examination revealed a "cambium-like" tumor zone and rhabdoid cells with high mitotic activity, strongly suggesting spindle cell ERMS. **IHC Results:** Tumor cells expressed only vimentin; specific myogenic markers (myogenin/MyoD1) were unavailable locally, preventing formal confirmation despite strong suspicion of ERMS. **Conclusion:** Accurate classification of pediatric ovarian mesenchymal tumors requires more than just morphological vigilance; it necessitates robust immunohistochemical capacity. Strengthening diagnostic infrastructure and regional pathology networks is essential to avoid inappropriate management and improve patient outcomes in resource-limited settings.

Keywords

Pediatric Ovarian Tumor, Spindle Cell Embryonal Rhabdomyosarcoma, Juvenile Fibrosarcoma, Immunohistochemistry, Differential Diagnosis

1. Introduction

Ovarian tumors in children are rare and pose major diagnostic and therapeutic challenges due to their histological diversity and often nonspecific clinical presentation. Their histopathological classification remains complex, making these lesions a critical issue in pediatric surgery [1]-[3]. The annual incidence of ovarian tumors in girls is estimated at 2.6 cases per 100,000. Among these, 10% - 20% are malignant, representing approximately 3% of all pediatric cancers in girls under 15 years of age [3]. According to the WHO classification, these tumors are grouped into three main categories based on their origin: epithelial tumors, germ cell tumors, and sex cord-stromal tumors [3] [4].

Beyond these classical entities, mesenchymal tumors, though exceptional, deserve special attention. Embryonal rhabdomyosarcoma, particularly its spindle cell variant, is a rare but diagnostically deceptive entity. This form, considered the most differentiated among rhabdomyosarcomas, occurs mainly in children under 10 years of age and can develop in the genitourinary tract, including the ovary [5]-[7].

Histologically, the spindle cell variant may mimic infantile fibrosarcoma due to its fascicular architecture, scant cytoplasm, and mild nuclear atypia. This morphological overlap increases the risk of diagnostic error, especially in the absence of immunohistochemistry. Indeed, infantile fibrosarcoma is generally negative for muscular markers (desmin, myogenin, MyoD1), unlike embryonal rhabdomyosarcoma, which expresses them variably but significantly [5]-[7].

We report here a case of spindle cell embryonal rhabdomyosarcoma of the ovary in a 7-year-old girl, initially misinterpreted as fibrosarcoma, in order to highlight the diagnostic difficulties encountered and the decisive role of immunohistochemistry in establishing the correct diagnosis.

2. Method

A retrospective study was conducted on a 7-year-old girl who presented with a right ovarian mass, initially diagnosed as juvenile fibrosarcoma, with a fatal outcome despite receiving three cycles of chemotherapy (Doxorubicin + Cisplatin) before the recommended immunohistochemical analysis was performed. The initial panel included only two markers: CD34 and S100.

For pedagogical purposes, we performed a morphological review which:

- Challenged the initial diagnosis, raising the possibility of spindle cell embryonal rhabdomyosarcoma;
- Consequently justified revision and expansion of the immunohistochemical panel to explore the newly suspected diagnosis. In addition to CD34 and S100, the following markers were added: myogenin, MyoD1, desmin, EMA, cytokeratin AE1/3, and vimentin.

3. Case Presentation

The patient, H.K., a 7-year-old girl with no significant medical history, was admitted to the pediatric department of the National Hospital of Zinder (NHZ) for abdominal pain associated with dysuria for about two weeks, without fever. On admission, her general condition was preserved, with normally colored mucous membranes. Clinical examination revealed a distended bladder; other systems were unremarkable. Bladder catheterization drained 400 mL of dark urine.

An etiological workup showed, on abdominopelvic ultrasound, a right ovarian mass with minimal ascites. Chest radiography and other investigations revealed no significant abnormalities. Exploratory laparotomy disclosed a right ovarian tumor invading the abdominal viscera and peritoneum. Complete tumor excision was performed, with an uneventful postoperative course.

Macroscopic findings: The ovarian mass measured 9 cm in its greatest dimension, weighed 200 g, and displayed friable areas with necrotic and hemorrhagic changes (Figure 1(a) and Figure 1(b)).

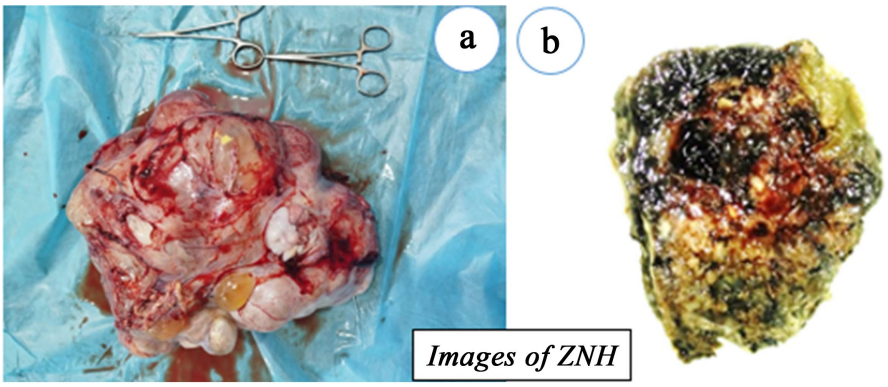


Figure 1. Macroscopic image of right ovarian mass (a: in the fresh state; b: in the fixed Stade).

Microscopic examination: Sections showed a malignant tumor proliferation

composed of infiltrative sheets of spindle-shaped cells (Figure 2(a)). The cells exhibited atypical nuclei with frequent mitotic figures, arranged in a "fishbone-like" fascicular pattern within a fibrous, edematous, and hemorrhagic stroma (Figure 2(a) and Figure 2(b)).

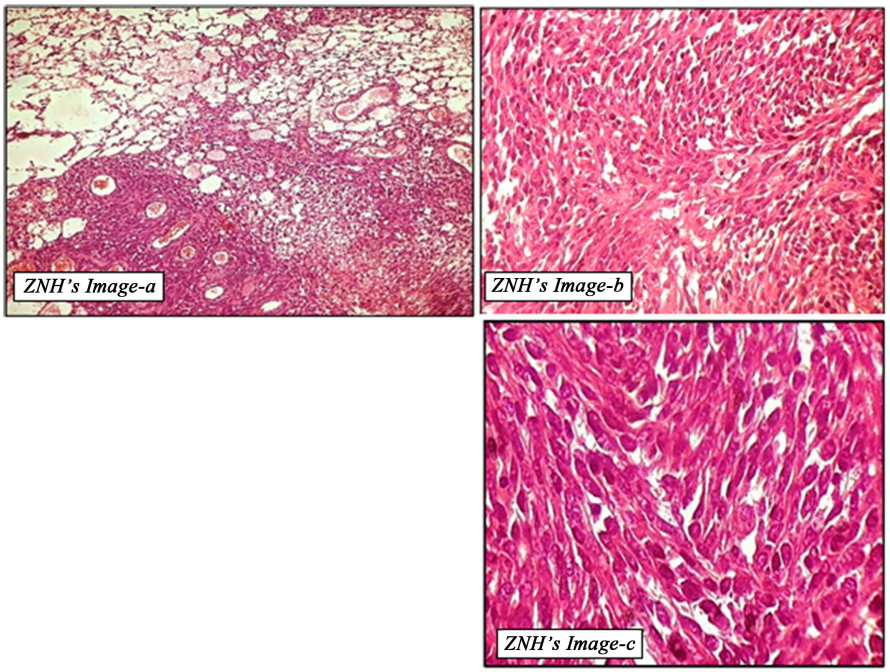


Figure 2. Microscopic images. a (HE ×100): Ovarian parenchyma showing areas of variable cellularity and a spindle cell proliferation. b (HE ×200) and c (HE ×400): Cellular areas with bundles of malignant spindle cells arranged in a "fishdone" pattern.

A diagnosis of juvenile fibrosarcoma was initially suggested, and immunohistochemical analysis was requested for CD34 and S100. Chemotherapy with Doxorubicin and Cisplatin was administered in three cycles. The patient died two weeks after the third cycle, before the immunohistochemistry could be performed.

Morphological review: The Re-examination revealed, in addition to features consistent with juvenile fibrosarcoma, a "cambium-like" tumor zone surrounded by markedly pleomorphic, hyperchromatic, and mitotically active cells. Clusters of rhabdoid cells were also noted, with abundant eosinophilic cytoplasm, eccentric vesicular nuclei, and frequent mitoses, suggesting high proliferative activity (Figure 3(a) and Figure 3(b)). These findings supported the diagnosis of spindle cell embryonal rhabdomyosarcoma and justified revising the immunohistochemical panel.

In addition to CD34 and S100 (previously tested), desmin, EMA, cytokeratin AE1/3, vimentin, myogenin, and MyoD1 (the latter two unavailable locally) were included. Immunostaining showed that only vimentin was expressed by the tumor cells (Figures 4(a)-(f)).

Based on the morphological review and immunohistochemical findings, the diagnosis of spindle cell embryonal rhabdomyosarcoma was strongly suspected, though not formally confirmed due to the unavailability of the specific myogenic

markers myogenin and MyoD1.

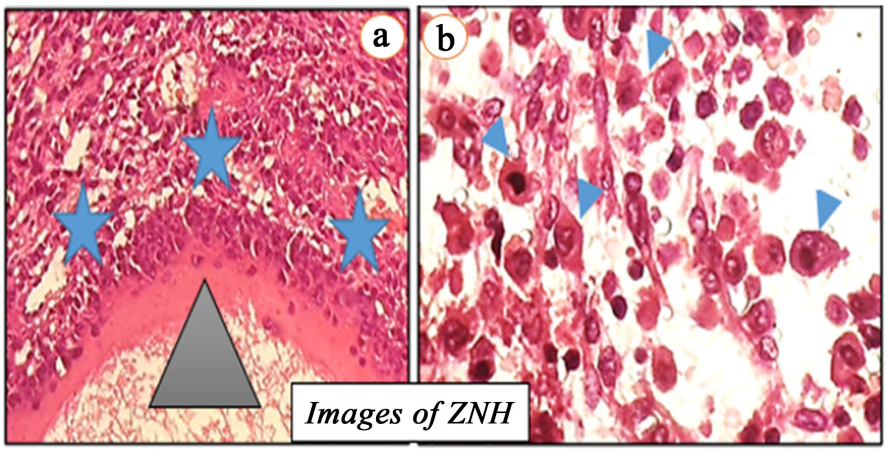


Figure 3. Microscopic images. a. (HE ×200): Central "Cambium-like" appearance [Wavy band of dense connective tissue (Gray triangle) surrounded by atypical cells arranged disorderly (Blue stars)]. b. (HE ×400): Cluster of rhabdoid cells (Arrowhead) in active mitosis (Eccentric nuclei, sometimes vesicular; Abundant eosinophilic cytoplasm).

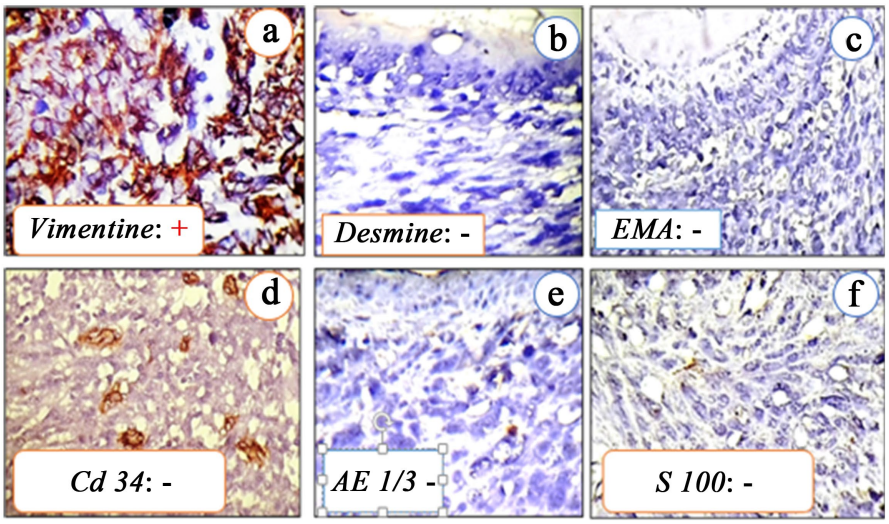


Figure 4. Immunohistochemical images of ZNH. Expanded immunohistochemical panel showing: Cytoplasmic expression of Vimentin; Lack of expression of the others antibodies (Desmin; CD 34; S100; EMA and EA 1/3).

4. Discussion

The case illustrates the diagnostic challenges posed by pediatric ovarian mesenchymal tumors, particularly in resource-limited settings. The initial diagnosis of juvenile fibrosarcoma was based solely on morphology, without immunohistochemical confirmation. Distinguishing histological features: spindle cell proliferation, poorly differentiated stroma, and absence of glandular or germ cell structures. So it illustrates the diagnostic ambiguity between juvenile fibrosarcoma and spindle cell ERMS. Both exhibit spindle morphology and overlapping histologic features [8] [9]. Spindle cell rhabdomyosarcomas may mimic fibroblastic or pe-

ripheral nerve-sheath tumors, but their high cellularity, atypia, and mitotic activity help distinguish them from benign proliferations. Immunohistochemistry is useful but not infallible: Desmin negativity does not entirely rule out ERMS, as antigen preservation depends on fixation quality and storage duration. False-negative Desmin staining has been reported in suboptimally processed specimens [10].

The diagnostic error had significant therapeutic consequences. But the choice of chemotherapy (Doxorubicin-Cisplatin) reflected financial limitations rather than diagnostic certainty. The standard VAC regimen (Vincristine, Actinomycin D, Cyclophosphamide) remains the recommended protocol for rhabdomyosarcoma [11]. The fatal outcome likely reflected advanced disease and limited supportive care, rather than delayed diagnosis alone.

Similar diagnostic pitfalls have been documented in pediatric ovarian rhabdomyosarcomas [12]-[15]. Consistent use of myogenic markers and improved access to IHC panels can reduce misclassification. Furthermore, continuous training for pathologists in morphological pattern recognition is vital.

Although the absence of immunohistochemistry contributed to the delay in diagnostic revision, it cannot be concluded that this alone caused the fatal outcome, as no autopsy was performed. The advanced tumor stage and poor clinical condition likely played decisive roles in prognosis. Even with an earlier rhabdomyosarcoma diagnosis, limited chemotherapy tolerance and inadequate supportive care might still have led to death. Nevertheless, this case highlights the importance of immunohistochemistry in accurately classifying pediatric soft-tissue tumors and guiding treatment. Expression of desmin, myogenin, and MyoD1 confirms rhabdomyoblastic differentiation, whereas fibrosarcoma typically expresses only vimentin [16]-[18]. Future reports should focus on correlations between tumor stage, pre- and post-treatment status, and therapy response to better understand outcomes in low-resource settings.

From an educational standpoint, this case underscores the importance of morphological vigilance, interdisciplinary collaboration, and investment in diagnostic infrastructure to ensure equitable cancer care in low-resource regions [19].

5. Conclusion

This case exemplifies the diagnostic challenges of pediatric ovarian mesenchymal tumors in resource-limited settings. The initial morphological misinterpretation, compounded by IHC unavailability, led to inappropriate management. Desmin negativity in this case likely reflected technical limitations rather than biological absence. Strengthening IHC capacity, promoting expert consultations, and developing regional pathology networks are essential for accurate tumor classification and better patient outcomes.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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