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Primary Retroperitoneal Mature Cystic Teratoma with Incidental Extramedullary Hematopoiesis in a 35-Year-Old Female

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Abstract

Primary retroperitoneal teratomas are rare germ cell tumors in adults. The presence of extramedullary hematopoiesis (EMH) within a benign mature cystic teratoma is an exceptionally scarce histopathological variant. We report the case of a 35-year-old female who presented with a large, well-defined retroperitoneal mass. Following successful complete surgical excision, histopathological examination revealed classic elements of a mature cystic teratoma interspersed with ectopic bone elements and foci of extramedullary hematopoiesis. The patient experienced an uneventful recovery with no signs of recurrence. This case emphasizes the diagnostic importance of comprehensive tissue sampling to rule out malignancy and identify unique benign tissue variations.

Keywords

Teratoma, Retroperitoneal, Extramedullary, Benign

1. Introduction

Teratomas are non-seminomatous germ cell neoplasms derived from totipotent cells that differentiate into tissues representing all three embryonic germ layers: ectoderm, mesoderm, and endoderm [1]. While they predominantly occur in the gonads (ovaries and testes), primary extragonadal teratomas are rare, accounting for less than 5% of all teratomas [1] [2]. The retroperitoneum is the third most

common site, representing 1% to 11% of primary retroperitoneal tumors, primarily seen in children and rarely in adults older than 30 years [1] [2].

Extramedullary hematopoiesis (EMH) refers to the production of blood cells outside the bone marrow, usually occurring secondary to severe hematological disorders like myelofibrosis, hemolytic anemias, or thalassemia [3]. However, its occurrence within a primary retroperitoneal mature cystic teratoma in an otherwise healthy patient is exceptionally rare [3]. We describe the clinical, surgical, and histopathological findings of this unusual composite entity.

2. Case Presentation

2.1. Clinical History and Initial Evaluation

A 35-year-old female presented to our urology department with a history of progressive, dull abdominal fullness and flank discomfort. She had no significant past medical history, no constitutional symptoms (fever, night sweats, or unexplained weight loss), and no known hematological anomalies.

Physical examination revealed a palpable, non-tender, relatively mobile mass in the abdomen felt occupying the left flank area on bimanual examination. Routine laboratory workup, including a complete blood count (CBC), liver function tests, renal function profiles, and tumor markers (beta-hCG, alpha-fetoprotein, and CEA), fell within normal reference ranges. Following the histopathological discovery of extramedullary hematopoiesis within the teratoma, a focused hematologic evaluation was performed to rule out systemic myeloproliferative diseases. Review of the patient's complete blood count (CBC) parameters was entirely unremarkable, and a peripheral blood smear demonstrated normal cellular morphology without circulating blasts or leucoerythroblastic features. A formal hematology consultation confirmed the absence of chronic hemolytic anemias or myelofibrosis, confirming that the hematopoietic proliferation was a localized, autonomous phenomenon within the tumor

2.2. Imaging and Pre-Operative Workup

An abdominal and pelvic computed tomography (CT) scan (**Figure 1**) revealed a large, well-circumscribed, encapsulated multi-locular retroperitoneal mass measuring approximately 23 × 17 × 10 cm. The mass was located in the retroperitoneal space, causing mild mass effect and displacing surrounding intra-abdominal structures anteriorly. The tumor demonstrated internal tissue heterogeneity, consisting of extensive fatty components, fluid-filled cystic zones, and isolated areas of dense calcification/ossification. There was no radiographical evidence of local infiltration into the major vessels, kidneys, or adrenal glands, and no distant metastasis was noted. Crucially, both ovaries, the uterus, and adjacent ipsilateral retroperitoneal organs were visualized as structurally normal and completely anatomically independent of the mass, excluding a gonadal or secondary adnexal origin.

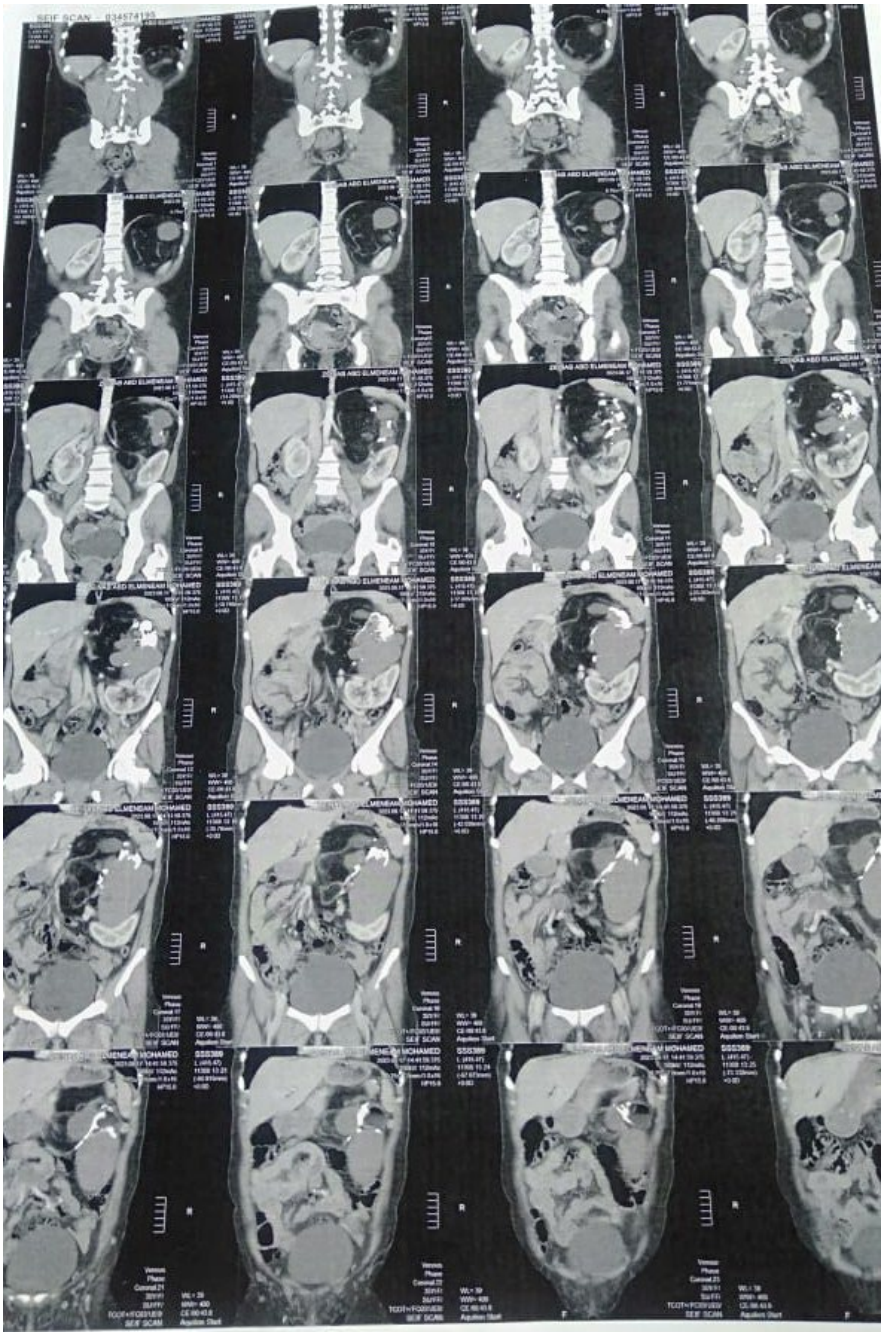


Figure 1. Pre-operative abdominal CT scan demonstrating a large, well-encapsulated, heterogeneous retroperitoneal mass with mixed adipose tissue, fluid attenuation, and foci of dense calcification.

3. Management and Surgical Intervention

Given the massive size and potential for local compressive complications, the patient underwent an elective surgical resection under general anesthesia. The patient was secured in the right lateral decubitus position, and a left supracostal flank incision was performed. The retroperitoneal space was entered sequentially by layers while maintaining meticulous intraoperative hemostasis.

Intraoperatively, a large, well-defined, encapsulated soft tissue mass was identified. The mass was successfully isolated and completely excised with negative macroscopic margins, without causing any intraoperative complications or measurable blood loss. Strict care was taken to maintain full capsular integrity to prevent the intraperitoneal spillage of highly irritative cystic contents. The total operative duration was approximately 180 minutes. Visual inspection during exploration confirmed that both ovaries and the surrounding viscera were entirely unaffected and free of disease. The patient experienced a smooth, uneventful postoperative recovery, was transferred to the regular inpatient ward, and was safely discharged home on postoperative day one.

The patient was placed on a regular surveillance protocol. At the 12-month postoperative follow-up interval, she remained completely asymptomatic. Recurrence was rigorously assessed via annual clinical examinations and follow-up contrast-enhanced abdominal/pelvic CT imaging, which demonstrated no evidence of local recurrence or residual disease.

4. Pathological Findings

4.1. Gross (Naked Eye) Examination

The pathology department received a well-defined, large soft tissue lesion measuring 23 × 17 × 10 cm. Sectioning of the mass demonstrated a highly heterogeneous structure composed of dense fibrofatty tissue, multi-loculated cystic cavities filled with thick, yellowish mucinous material, and interspersed hard, calcified/bony focus areas. To guarantee a comprehensive diagnostic evaluation and definitively rule out hidden immature or malignant components, an exhaustive gross sampling protocol was implemented. A total of 15 formal tissue blocks were harvested from all grossly distinct solid, cystic, calcified, and adipose zones for thorough microscopic review. Serial sectioning of these blocks confirmed the complete absence of atypical cells, immature neuroepithelium, or somatic malignant transformations.

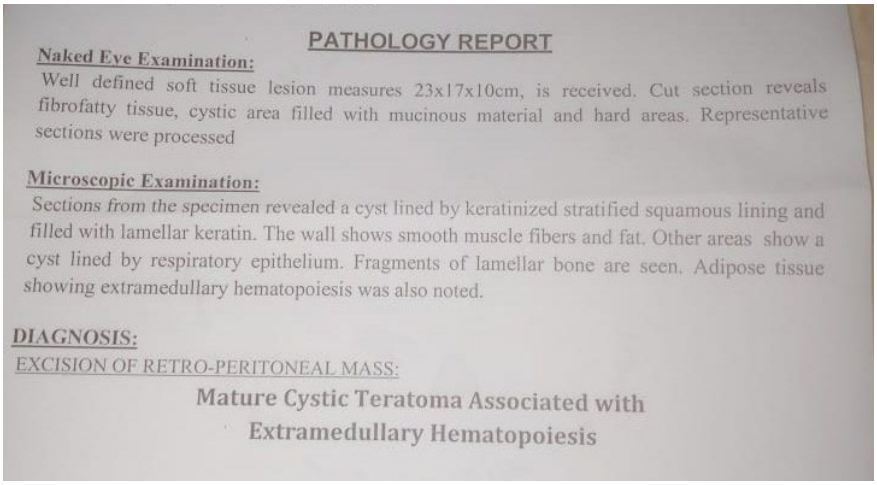
4.2. Microscopic Examination

Histopathological analysis of the stained sections revealed distinct mature components representing multiple germ layers:

- **Ectodermal Derivatives:** Cystic cavities lined by mature, keratinized stratified squamous epithelium filled with an abundance of lamellar keratin flakes.
- **Mesodermal Derivatives:** The cyst walls demonstrated organized smooth muscle fibers, mature adipose (fat) tissue, and distinct fragments of mature lamellar bone.
- **Endodermal Derivatives:** Distinct focal cystic areas were found lined by mature, pseudostratified ciliated respiratory epithelium.

Crucially, interspersed within the mature adipose tissue, dense nests of active extramedullary hematopoiesis (EMH) were noted, characterized by the presence of a polymorphic population of hematopoietic cells, including erythroid precur-

sors, myeloid elements, and megakaryocytes. No cytological atypia, immature neuroepithelium, or malignant transformations were observed, confirming a definitive diagnosis of a Mature Cystic Teratoma Associated with Extramedullary Hematopoiesis (Figure 2).


PATHOLOGY REPORT

Naked Eye Examination:

Well defined soft tissue lesion measures 23x17x10cm, is received. Cut section reveals fibrofatty tissue, cystic area filled with mucinous material and hard areas. Representative sections were processed

Microscopic Examination:

Sections from the specimen revealed a cyst lined by keratinized stratified squamous lining and filled with lamellar keratin. The wall shows smooth muscle fibers and fat. Other areas show a cyst lined by respiratory epithelium. Fragments of lamellar bone are seen. Adipose tissue showing extramedullary hematopoiesis was also noted.

DIAGNOSIS:

EXCISION OF RETRO-PERITONEAL MASS:

Mature Cystic Teratoma Associated with Extramedullary Hematopoiesis

Figure 2. Histopathological examination showing mature adipose tissue and lamellar bone fragments interspersed with active hematopoietic nests containing erythroid, myeloid, and megakaryocytic lineages (H & E stain).

5. Discussion

Primary retroperitoneal teratomas are exceptionally rare germ cell tumors in adults, representing less than 11% of all primary retroperitoneal neoplasms [1] [2]. Unlike their gonadal counterparts, primary extragonadal retroperitoneal teratomas originate from displaced primordial germ cells that fail to complete their migration along the urogenital ridge during early embryogenesis [1]. In adults, these tumors exhibit an indolent growth pattern and remain clinically silent for extended periods. They are frequently discovered incidentally or only after attaining a massive size, at which point they manifest with non-specific symptoms such as abdominal fullness, dull flank pain, or palpable abdominal masses caused by the compression of adjacent retroperitoneal viscera [1] [2].

Radiographically, computed tomography (CT) and magnetic resonance imaging (MRI) serve as the cornerstones for pre-operative evaluation. The presence of a well-encapsulated, heterogeneous retroperitoneal mass demonstrating a clear coexistence of adipose tissue, fluid-attenuating cystic spaces, and coarse, dense calcifications or ossifications is highly pathognomonic for a mature cystic teratoma [4]. However, when retroperitoneal masses present with mixed fatty and hematopoietic components, they can easily mimic other distinct clinical entities. These include primary retroperitoneal liposarcomas, exophytic developmental duplication cysts, or retroperitoneal myelolipomas [4].

The most unique and physiologically remarkable feature of this case is the microscopic identification of active extramedullary hematopoiesis (EMH) interspersed within the teratoma's mature adipose tissue matrix. Clinically, EMH func-

tions as a systemic, compensatory physiological response to bone marrow insufficiency, typically triggered by underlying hematological disorders such as myelofibrosis, severe hemolytic anemias, hemoglobinopathies, or marrow-infiltrating neoplastic processes [3]. However, the development of localized EMH within a primary extragonadal teratoma in a patient with completely normal peripheral blood profiles and no systemic hematological diseases represents a distinct, autonomous localized phenomenon [3].

This localized hematopoietic proliferation can be explained by two prevailing scientific theories:

1) The Bone Marrow Niche Theory: The close structural and spatial alignment of mature lamellar bone fragments and adjacent fatty adipose stroma within the teratoma essentially recapitulates a functional, microscopic bone marrow niche [3]. This unique osteo-adipose microenvironment possesses the precise architectural and biochemical properties required to home circulating pluripotent mesenchymal cells or migrating CD34+ hematopoietic stem cells, signaling them to settle, proliferate, and differentiate into mature erythroid, myeloid, and megakaryocytic lineages [3].

2) Totipotent Germ Cell Differentiation: Because teratomas arise from totipotent germ cells capable of differentiating into any tissue of the three embryonic germ layers, the hematopoietic nests may simply reflect a direct, highly specialized mesodermal differentiation pathway where the tumor independently generates its own stromal and hematological architecture.

From a therapeutic perspective, complete surgical excision via exploratory laparotomy or a minimally invasive approach remains the definitive standard of care [1] [5]. Total capsular resection minimizes the risk of local recurrence and prevents significant chemical peritonitis or acute inflammatory responses that can be triggered by the intra-abdominal spillage of highly irritative, keratinaceous, or mucinous cystic fluids [5]. Furthermore, securing the entire, intact surgical specimen is vital for a comprehensive histopathological evaluation. Pathologists must implement an exhaustive tissue sampling and sectioning protocol to meticulously scan for any focal micro-regions containing immature embryonic tissue (specifically neuroepithelium) or somatic malignant transformations, such as squamous cell carcinoma or adenocarcinoma arising within the benign components [6]. The identification of these elements entirely alters the staging, prognosis, and postoperative oncological management of the patient.

Ethics Approval and Consent to Participate

The institutional review board or local ethics committee provides a waiver of approval for single retrospective case reports that do not involve experimental interventions.

Consent for Publication

Written informed consent was obtained from the patient for the publication of

this case report and any accompanying intraoperative and radiological images. A copy of the written consent form is available for review by the Editor-in-Chief of this journal.

Conflicts of Interest

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

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