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Case Report

Paraneoplastic Nephrotic Syndrome in a Teenager with Ovarian Teratoma: Successful Treatment with Tumor Resection and Ovarian Tissue Preservation

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ABSTRACT

Nephrotic syndrome is typically associated with primary glomerular diseases but can occasionally manifest as a paraneoplastic phenomenon. We present the case of a 14-year-old girl who developed nephrotic syndrome accompanied by significantly elevated CA 19-9 levels. She was subsequently found to have a 7 cm mature ovarian teratoma. Despite a non-diagnostic renal biopsy, imaging studies and tumor marker analyses guided the decision to surgically remove the tumor while preserving ovarian tissue. Postoperatively, her nephrotic syndrome resolved completely, and CA 19-9 levels normalized within a month. This case underscores the importance of considering paraneoplastic causes in unexplained nephrotic syndrome and highlights the critical role of timely diagnosis and a multidisciplinary approach in achieving full recovery.

Keywords: Paraneoplastic nephrotic syndrome; Ovarian teratoma; Pediatric nephrotic syndrome; Renal biopsy; Tumor resection; Remission

Introduction

Paraneoplastic phenomena represent an intriguing interplay between tumors and systemic manifestations, often complicating diagnostic and therapeutic pathways. In nephrotic syndrome, these associations can manifest as immune-mediated injuries to the glomeruli, triggered by secreted tumor products or immune responses. Understanding these rare connections is essential for timely intervention and improved outcomes[1].

Nephrotic syndrome, commonly linked to primary glomerular diseases, can occasionally arise as a paraneoplastic phenomenon, particularly in the presence of systemic conditions such as neoplasms[1,2]. While rare in pediatric patients, identifying paraneoplastic nephrotic syndrome is vital, especially when tumor markers are elevated without an apparent cause[3].

This report describes a 14-year-old girl diagnosed with nephrotic syndrome associated with a mature ovarian teratoma. The case highlights the diagnostic challenges and clinical importance of recognizing paraneoplastic nephrotic syndrome, particularly in cases with unexplained etiology and elevated tumor markers. It also underscores the value of a multidisciplinary approach that integrates clinical, imaging, and pathological evaluations for early diagnosis and effective management.

Case Report

A 14-year-old female presented with periorbital and lower limb edema. Laboratory evaluations revealed significant proteinuria, hypoalbuminemia, and hyperlipidemia, leading to a diagnosis of nephrotic syndrome. Renal biopsy was inconclusive[1]. Further evaluation, including abdominal ultrasound and MRI, revealed

a 7 cm mass in the right ovary with benign imaging features **(Figure 1)**. Elevated serum CA 19-9 levels were detected, while other tumor markers remained within normal limits.

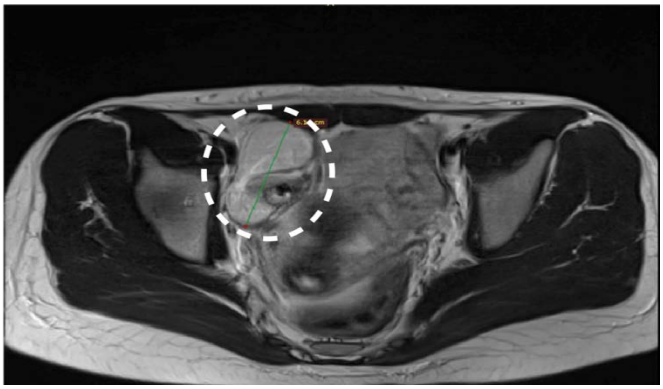

Figure 1: Axial Magnetic Resonance Imaging (MRI) reveals a well-defined ovarian mass in the right adnexa, which is consistent with a mature ovarian teratoma. The lesion exhibits benign imaging characteristics without evidence of invasion.

The patient underwent surgery to excise the right ovarian tumor via a Pfannenstiel incision, preserving the adjacent normal ovarian parenchyma. Peritoneal fluid samples obtained pre- and post-excision, as well as after peritoneal lavage, showed no evidence of neoplastic cells **(Figure 2)**.

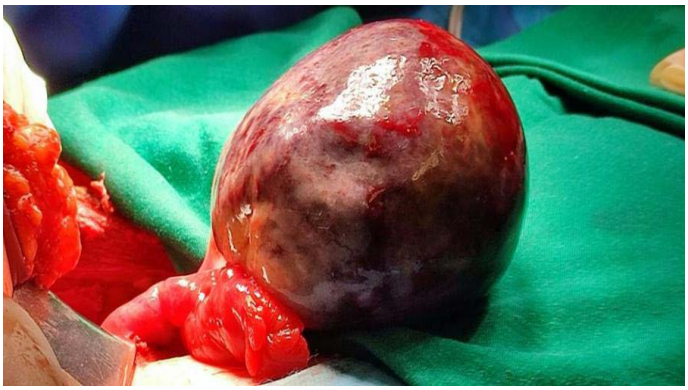

Figure 2: Intraoperative image of the resected mature ovarian teratoma, demonstrating its encapsulated and well-circumscribed appearance. The tumor was surgically removed with preservation of adjacent ovarian tissue.

Postoperatively, the patient achieved complete remission of nephrotic syndrome, which has remained stable for one year. Histopathological examination confirmed the diagnosis of a mature ovarian teratoma. Serum CA 19-9 levels normalized within one month of tumor resection[3].

The histopathological findings provided essential confirmation of the diagnosis, supporting the benign nature of the mature ovarian teratoma. No evidence of malignancy or metastatic cells was observed in the intraoperative cytological evaluations of peritoneal fluid samples. These results validated the hypothesis of a paraneoplastic mechanism and reinforced the decision to preserve adjacent ovarian tissue, which is critical for maintaining long-term reproductive health in young patients. Furthermore, the benign nature of the tumor underscores its systemic effects as being immune-mediated rather than directly invasive.

Discussion

This case demonstrates the rare association of nephrotic syndrome with an ovarian teratoma as a paraneoplastic phenomenon. Such occurrences are rare in pediatric patients and require a high index of suspicion for timely diagnosis and treatment[4,5].

Paraneoplastic nephrotic syndrome is often attributed to tumor-secreted products, including cytokines, growth factors, or antigens, which induce immune-mediated glomerular injury. In this patient, the elevated CA 19-9 levels and the presence of an ovarian teratoma suggest that the tumor triggered an immune response, resulting in glomerular damage[3]. While mature teratomas are generally benign, their potential to elicit systemic effects underscores the complexity of their biological activity[4].

Diagnosing paraneoplastic nephrotic syndrome is challenging due to its rarity and the absence of specific markers linking the tumor to nephrotic syndrome. The utility of renal biopsy in such cases is debated, as histological findings often fail to distinguish primary nephrotic syndrome from paraneoplastic variants[1,2]. In this case, the non-diagnostic renal biopsy, along with the identification of a benign ovarian tumor and elevated tumor markers, was critical in diagnosing the paraneoplastic condition.

This case highlights the importance of thorough evaluation in nephrotic syndrome of unexplained origin, particularly in the presence of elevated tumor markers. Imaging studies and tumor marker analysis should be integral to the diagnostic process. Early recognition and surgical excision of the underlying tumor can lead to complete remission of nephrotic syndrome, as demonstrated here[2]. Preservation of ovarian tissue during surgery was crucial for the patient's long-term reproductive health, especially given her young age[4,5].

Although paraneoplastic nephrotic syndrome has been reported in association with various malignancies, including Hodgkin's lymphoma and renal cell carcinoma, its occurrence with benign ovarian teratomas is exceptionally rare[4,5]. The limited cases described in the literature underscore the potential for benign tumors to cause significant systemic effects through paraneoplastic pathways. Further research and clinical studies are necessary to better understand its pathogenesis and establish reliable diagnostic markers[5].

Conclusion

This case underscores the importance of evaluating potential neoplasms in nephrotic syndrome of unexplained origin, particularly in the presence of elevated tumor markers. The complete resolution of nephrotic syndrome following excision of a mature ovarian teratoma confirms the tumor's role in this paraneoplastic phenomenon, serving as a diagnostic criterion. The role of renal biopsy remains a subject of debate, requiring individualized consideration based on clinical findings. Early diagnosis and multidisciplinary management are critical for optimal outcomes, as demonstrated in this case, where timely identification and surgical treatment led to full recovery[2,4,5]. This case reinforces the need to consider paraneoplastic causes in complex nephrotic syndrome presentations, given their potential for significant systemic impact and reversibility with appropriate treatment.

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