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An unusual pseudotumor cerebri syndrome case report featuring a massive retroperitoneal cyst

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SUMMARY

Pseudotumor Cerebri Syndrome (PTCS), also known as idiopathic intracranial hypertension, is a rare condition characterized by increased intracranial pressure without identifiable structural or biochemical causes. Herein, we present a unique case of PTCS in conjunction with a massive retroperitoneal cyst, an exceptionally rare co-occurrence. This case highlights the importance of considering a typical presentations and conducting thorough investigations to guide appropriate management strategies. Pseudotumor Cerebri Syndrome (PTCS) is a relatively uncommon disorder characterized by Increased Intracranial Pressure (ICP) without any underlying structural abnormalities or identifiable etiology. Patients typically present with symptoms such as headache, visual disturbances, and pulsatile tinnitus. The diagnosis of PTCS is often challenging due to its diverse clinical manifestations and the absence of specific diagnostic markers.

Keywords: Pseudotumor; Etiology; Intracranial; Biochemical; Cyst; Investigations; Pressure

INTRODUCTION

We report the case of a 42-year-old female who presented to our neurology clinic with complaints of persistent headaches, blurred vision, and intermittent dizziness over the past six months. She had no significant medical history, including no prior history of head trauma or neurosurgical interventions. Physical examination revealed papilledema bilaterally and mild visual field deficits on formal perimetry. Neuroimaging studies, including Magnetic Resonance Imaging (MRI) of the brain and Computed Tomography (CT) of the head, were unremarkable, with no evidence of mass lesions or hydrocephalus. Given the clinical suspicion of PTCS, a lumbar puncture was performed, revealing an elevated opening pressure of 35 cm H2O. Cerebrospinal Fluid (CSF) analysis showed normal cell counts and chemistry. The patient was diagnosed with PTCS and initiated on acetazolamide therapy for intracranial pressure management.

However, the patient's symptoms persisted despite treatment, prompting further investigations. Abdominal ultrasound was performed, revealing a large retroperitoneal cyst measuring approximately 15 cm in diameter, displacing adjacent organs. Subsequent abdominal MRI confirmed the presence of the cyst, which appeared to arise from the left renal capsule. The coexistence of PTCS with a massive retroperitoneal cyst in our patient represents a rare and unusual presentation. Retroperitoneal cysts are typically benign lesions arising from various structures, including the kidneys, adrenal glands, and lymphatic system. While retroperitoneal cysts are often asymptomatic and discovered incidentally on imaging studies, they can occasionally cause symptoms such as abdominal pain, distension, or compression of adjacent structures [1].

LITERATURE REVIEW

The pathophysiological relationship between PTCS and retroperitoneal cysts remains unclear. It is plausible that the retroperitoneal cyst in our patient may have contributed to the elevation of intracranial pressure through mechanisms such as venous congestion or impaired cerebrospinal fluid absorption. Alternatively, the coexistence of PTCS and the retroperitoneal cyst may be coincidental, with each condition representing independent pathological processes. Management of our patient posed several challenges due to the presence of concurrent PTCS and the retroperitoneal cyst. Treatment strategies aimed to alleviate symptoms and reduce intracranial pressure while addressing the underlying

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etiology of the retroperitoneal cyst. Surgical intervention was considered for the retroperitoneal cyst, given its significant size and symptomatic nature. However, the risks and benefits of surgical excision needed to be carefully weighed against the potential complications, including injury to adjacent structures and recurrence of the cyst [2].

DISCUSSION

We present a rare case of pseudotumor cerebri syndrome occurring concurrently with a massive retroperitoneal cyst, highlighting the importance of considering atypical presentations and conducting thorough investigations in patients with intracranial hypertension. Further research is warranted to elucidate the pathophysiological mechanisms underlying this unusual association and to optimize management strategies for such complex cases. The simultaneous occurrence of PTCS and a massive retroperitoneal cyst in our patient represents an unusual clinical scenario. While PTCS is typically associated with neurological symptoms related to elevated ICP, the presence of a retroperitoneal cyst introduces an additional diagnostic challenge. Retroperitoneal cysts are rare entities, with diverse etiologies including developmental, traumatic, neoplastic, and infectious causes [3].

In this case, the retroperitoneal cyst was asymptomatic and incidentally discovered during the evaluation of the patient's gastrointestinal symptoms. Its association with PTCS remains speculative, as there is limited literature linking retroperitoneal cysts to elevated ICP. However, given the absence of other identifiable causes for PTCS in this patient, the retroperitoneal cyst warrants consideration as a potential contributing factor. The management of this patient posed several challenges. Treatment strategies for PTCS typically aim to reduce ICP through weight loss, dietary modification, medications (such as acetazolamide), and, in refractory cases, surgical interventions such as optic nerve sheath fenestration or Cerebrospinal Fluid (CSF) shunting. However, the presence of a retroperitoneal cyst added complexity to the case, raising concerns about the need for surgical intervention and its potential impact on ICP dynamics [4-6].

CONCLUSION

Ultimately, a multidisciplinary approach involving neurology, neurosurgery, and general surgery was adopted to address both the PTCS and the retroperitoneal cyst. The patient was initiated on acetazolamide therapy for PTCS and underwent elective surgical resection of the retroperitoneal cyst to alleviate potential mass effect and mitigate the risk of complications such as cyst rupture or hemorrhage. We report a rare case of PTCS coexisting with a massive retroperitoneal cyst, highlighting the importance of considering atypical presentations and conducting thorough investigations to uncover potential underlying causes. This case underscores the complexity of managing concurrent medical conditions and the necessity of a multidisciplinary approach to optimize patient outcomes. Further research is warranted to elucidate the potential association between PTCS and retroperitoneal cysts, which may contribute to a better understanding of the pathophysiology of both conditions.

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CONFLICT OF INTEREST

None.

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