CASE REPORT

Idiopathic Retroperitoneal Fibrosis with Bilateral Varicocele – A Rare Presentation of Very Rare Clinical Entity

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Abstract:

Retroperitoneal fibrosis is a very rare inflammatory disease leading to extensive fibrosis throughout the retroperitoneum. The majority of cases are idiopathic in which the exact cause is not known. The clinical presentation in retroperitoneal fibrosis is vague and nonspecific, and therefore the diagnosis relies mainly on radiological findings. Here we describe a rare case of idiopathic retroperitoneal fibrosis resulting in Grade 3 varicocele formation. To the best of our knowledge this is the first case report of its kind.

Keywords: Retroperitoneal Fibrosis, Idiopathic, Varicocele

Introduction:

Retroperitoneal Fibrosis (RPF) is a very rare inflammatory disease leading to extensive fibrosis throughout the retroperitoneum. Majority of cases are idiopathic accounting for more than 70% [1, 2]. The clinical presentation in RPF is vague and nonspecific, and therefore the diagnosis relies mainly on imaging studies. A diagnosis of idiopathic RPF can be made by using Computed Tomography (CT) scan or Magnetic Resonance Imaging (MRI) [1, 2]. Histological confirmation may sometimes be needed in doubtful cases. Recently, we came across a case of idiopathic RPF resulting in Grade 3 bilateral varicocele formation. To the best of our knowledge, a case of idiopathic RPF resulting in a Grade 3 varicocele has not been reported so far and this is the first report of its kind.

Case Report:

A 50 year-old male presented with pain in abdomen and back, and swelling in left groin associated with intermittent mild pain. The pain was insidious in onset and gradually progressive. The pain was dull aching in nature and increased in intensity after consumption of food. He also gave history of high grade fever one week back associated with chills and rigor. There was also history of gradual weight loss and loss of appetite. There were no other co-morbidities. He had undergone right hernioplasty 12 years back and left hernioplasty 4 years back. Physical examination revealed tenderness at left lumbar and iliac regions with no palpable mass or organomegaly. There was a single reducible swelling above left groin crease extending into left scrotum. Swelling was mildly tender without signs of inflammation. Cough impulse was absent. Abdomino-pelvic CT scanning revealed ill defined soft tissue density lesions at the retroperitoneal region with engulfment of the aorta and inferior vena cava. Bilateral varicocele was also noted with more on the left side and bulky left spermatic cord with loss of surrounding fat planes. These findings revealed a diagnosis of retroperitoneal fibrosis with evidence of Grade 3 varicocele formation.

Discussion:

RPF is a rare disease. Majority of the cases are idiopathic accounting for more than 70%. The cases of RPF show varied clinical presentations with vague and non-specific symptoms making the diagnosis difficult and thereby causing delay in the diagnosis. The diagnosis is made primarily on the basis of imaging studies. CT and MRI have become mainstays in the diagnosis and allow comprehensive evaluation of the extent and sequelae of the disease [1, 2]. At present spiral CT of the abdomen and pelvis without the administration of contrast is the most common method used for diagnosis.

Cases of RPF with varied clinical presentations have been reported previously [3-5], but the

present case is very unusual and rare because of its association with Grade 3 varicocele. To the best of our knowledge, it is the first case report of idiopathic RPF resulting in Grade 3 varicocele formation, which had never been reported in the past.

Conclusion:

The results show that as reported earlier clinical presentation with vague and non-specific symptoms making the diagnosis difficult is very common feature of RPF. This case also suggests that varicocele formation may be one of the varied clinical presentations.

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