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Bladder Wall Plexiform Neurofibromatosis in A Patient with NF1: Case Report and Literature Review

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Abstract

While von Recklinghausen provided a detailed description of the clinical entity of neurofibromatosis and is credited with its discovery, Smith initially described the condition in 1849. With an estimated frequency of one in every 3000 live births, it is a hereditary dysgenesis transmitted as a Mendelian dominant trait. It is a main neural crest derivation condition that also affects the nerve sheath's fibroblasts, Schwann cells, and supportive mesenchymal elements. Nerves of the pelvic, vesical, and prostatic plexuses have been demonstrated to be the source of neurofibromas of the genitourinary tract. The most often affected organ in the urinary tract is the bladder, which can develop as a diffuse infiltrative process as shown in the instances reported in this study, or as an isolated mass.

Introduction

While von Recklinghausen provided a detailed description of the clinical entity of neurofibromatosis and is credited with its discovery, Smith initially described the condition in 1849. With an estimated frequency of one in every 3000 live births, it is a hereditary dysgenesis transmitted as a Mendelian dominant trait. It is a main neural crest derivation condition that also affects the nerve sheath's fibroblasts, Schwann cells, and supportive mesenchymal elements. Nerves of the pelvic, vesical, and prostatic plexuses have been demonstrated to be the source of neurofibromas of the genitourinary tract. The most often affected organ in the urinary tract is the bladder, which can develop as a diffuse infiltrative process as shown in the instances reported in this study, or as an isolated mass. Is the elimination of When a fundamentally benign tumor necessitates the simultaneous extraction of pelvic organs or results in nerve damage, is it necessary or justified? We go over the best management practices and the literature. When a tumor of this kind is essentially benign and necessitates the simultaneous removal of pelvic organs or results in nerve damage, is its removal appropriate or necessary? We go over the best management practices and the literature.

Case report

In addition to lower abdominal pain, constipation, more than six café au lait patches on her torso, many subcutaneous nodules, and a palpable lower abdominal mass originating from her pelvis, the 19-year-old female patient also presented with hesitation, urgency, and frequency. An ultrasound revealed a heterogeneous, primarily hypoechoic mass bordering the bladder and prevertebral region that emerged from the bladder's posterior wall. An intravenous urogram (IVU) revealed a mass encasing the bilateral ureter in the lower third,although the top two thirds of the ureters were normal.

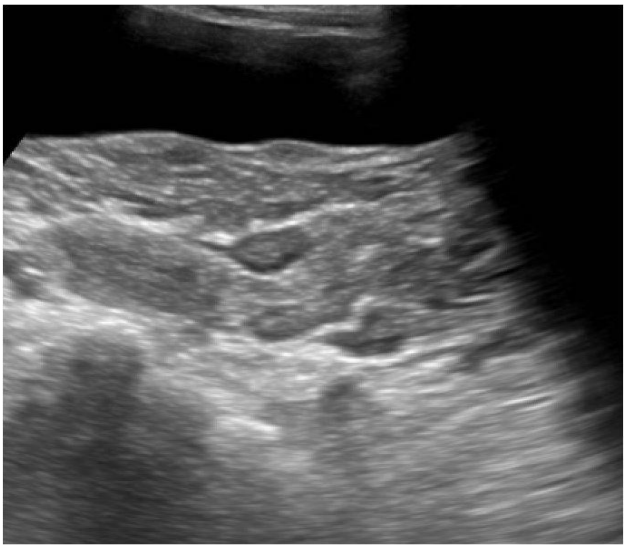

Figure:1-USG sagittal view of bladder showing predominantly hypoechoic lesion arising from posterior bladder wall.

Figure2: CT IVU delayed phase sagittal image. Contrast filled the bladder. Iso to hypodense lesion arising from post-bladder wall

A sizable irregularly lobulated heterogeneously enhancing soft tissue mass situated between the bladder and uterus was discovered by a contrast-enhanced computed tomography (CECT) scan. The mass had moved the bladder, invaded deep into the gluteus maximus, and encased the rectum, anus, and perianal region after passing through the larger sciatic notch and between the ischium and sacrum.

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To identify upper tract blockage, which could indicate a tumor's progressive growth and potential malignant transformation, careful monitoring is required. If the tumor grows larger, it should be removed.

Figure:3-CECT sagittal image. Heterogeneously enhancing mass lesion.

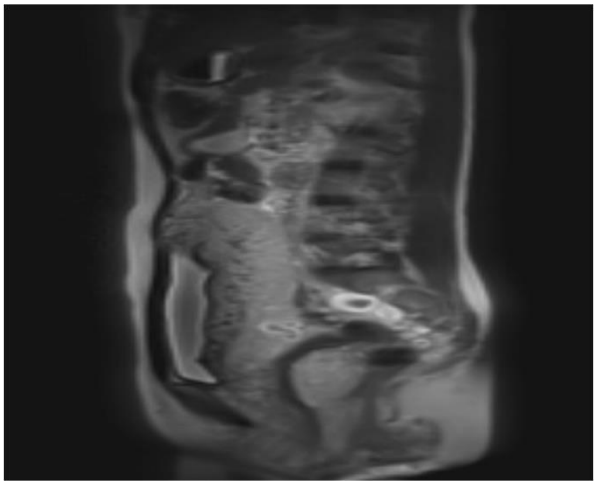

Figure:4-sagittal T2W image, Predominantly Hyperintense lesion.

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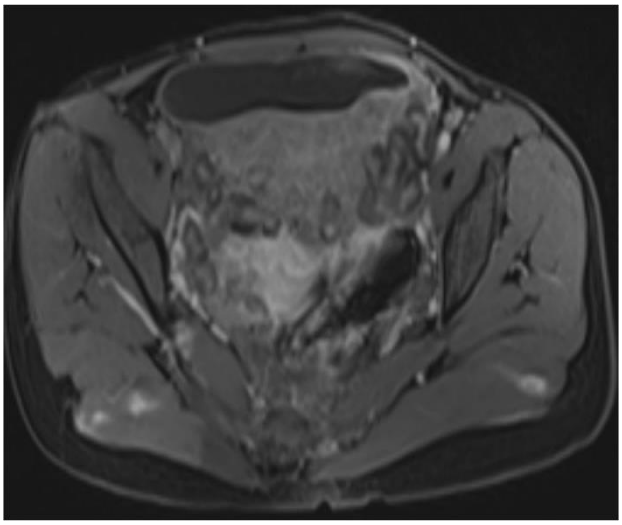

Figure:5-T1 post contrast. Heterogeneously enhancing mass lesion.

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