Spontaneous Vesicovaginal Fistula in Neurofibromatosis: A Case Report

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Abstract

Background: Neurofibromatosis type 1 is an autosomal dominant transmitted disease with various forms of clinical presentation. It commonly affects the skin and the nervous system. Involvement of the genitourinary system is rare. The bladder is the most commonly affected organ in the urinary system. Vesicovaginal fistula presenting as continuous leakage of urine per vagina is a rare urogenital manifestation of neurofibromatosis which has not been previously reported to the best of our knowledge. Vesicovaginal fistula is not a usual complication of urethral catheterization. Our aim is to report a rare case of vesicovaginal fistula in a patient with neurofibromatosis.

Case presentation: We report a case of a 36 year-old nullipara with eighteen years history of multiple features of type 1 neurofibromatosis who presented with continuous leakage of urine per vagina of spontaneous onset. Patient had urinary incontinence following which she was catheterized for 18 years but subsequently started leaking urine despite catheterization five months prior to presentation. There were no other known associated predisposing factors. Important findings on examination were paraplegia, multiple neurofibroma, café-au-lait spot and bony deformities including scoliosis. Examination in theatre showed vesicovaginal (juxta-urethral) fistula. Abdominal computerized tomography scan showed left diaphragmatic crural cyst, dextroscoliosis and cholelithiasis. Pelvic CT scan revealed irregular thickening of the bladder wall, uterine leiomyomata and left hemi pelvic deformity/dysplasia with resultant hip dislocation suggestive of neurofibromatosis of bone. Chest x-ray noted a soft tissue mass in the posterior mediastinum. She had surgical repair of vesicovaginal fistula using the vaginal approach.

Conclusion: The cause of vesicovaginal fistula in this patient with neurofibromatosis is uncertain. However, it may have resulted from bladder neurofibromatosis or prolonged urethral catheterization or both.

Keywords: Neurofibromatosis type 1; Urethral catheterization; Vesicovaginal fistula

Introduction

Neurofibromatosis type 1 or Von Recklinghausen's disease is an autosomal dominant transmitted disease which is located on chromosome 17q11 with various clinical manifestations [1,2]. The incidence of systemic disease is 1 in every 3000 births [3]. Usually, fifty percent of cases are sporadic, with no family history of the disease [4,5]. Skin is the most commonly affected site in neurofibromatosis type 1, [5] however any organ may be affected. Important features of this disease include hyperpigmented skin lesions (café-au-lait spots), neurofibromas, iris haematomas, macrocephaly, central nervous system tumours, defects of the skull and facial bones and vascular lesions [3-7]. Neurofibromatosis type 1 is quite distinct from neurofibromatosis type 2 which mainly causes vestibular schwannoma, presenting as progressive hearing loss [4,8].

Genitourinary tract is rarely involved in neurofibromatosis type 1, however the bladder is the commonest organ affected in the urinary tract and a few cases have been reported [3,9-12]. Neurofibromas of the genital tract are commonly found in the vulva, clitoris and labia but rarely found in vagina, cervix, endometrium and myometrium [13-15]. Bladder neurofibroma may present as a diffuse infiltrative process or an isolated neurofibroma. The symptoms vary, ranging from urinary incontinence to retention [10,14,15]. We are not aware to the best of our knowledge of any case of vesicovaginal fistula that resulted from neurofibromatosis with bladder involvement. Vesicovaginal fistula is also not a usual complication of urethral catheterization.

We present a rare case of vesicovaginal fistula in a patient with multiple features of neurofibromatosis.

Case presentation

A 36 year-old nulliparous lady with a history of involuntary leakage of urine, paraesthesia of lower limbs, leg weakness and spasms involving both legs and multiple nodules on the skin of 18 years duration. She presented with involuntary leakage of urine of 5 months duration despite urethral catheterization. She was diagnosed with neurofibromatosis 18 years prior to presentation and had excision of lump from her back following a history of paraesthesia of both lower limbs with associated weakness. She subsequently became unable to walk and developed urinary incontinence. Urinary incontinence resolved with continuous urethral catheterization using Foley’s catheter that was changed on a two weekly basis and inflated with 20 ml of fluid. However, five months prior to presentation she developed urinary incontinence despite catheterization. There were no other positive findings from the history.

On examination, she was paraplegic and had multiple neurofibroma all over her body with the largest measuring 3 cm × 3 cm × 3 cm. This is shown in Figure 1. She also had café-au-lait spot on her back measuring 4 cm × 2 cm. She had bony deformities involving both toes and scoliosis. Examination in theatre showed that the vulva was wet,
with continuous leakage of urine and dye test was positive. A juxta-urethral fistula was noted (shown in Figure 2), measuring 2 cm x 1 cm. Cough impulse was negative. A clinical diagnosis of juxta-urethral fistula in a patient with neurofibromatosis was made.

Figure 1: Multiple Neurofibromas in our patient.

Computerized tomography of the abdomen showed a cystic mass posterior to the spleen and expanding a section of the diaphragmatic crus (suggestive of left diaphragmatic crus cyst) and multiple hyperdense calculi in the gall bladder (cholelithiasis). Computerized tomography of the pelvis showed a cystic mass in a patient with neurofibromatosis in the gall bladder (cholelithiasis). Computerized tomography of the pelvis showed a cystic mass in a patient with neurofibromatosis in the gall bladder (cholelithiasis). Computerized tomography of the pelvis showed a cystic mass in a patient with neurofibromatosis in the gall bladder (cholelithiasis).

Discussion

Neurofibromatosis type 1 is an autosomal dominant disease with wide variability of clinical forms [1,2]. It predominantly affects the skin and nervous system [1,5]. The diagnosis can be made if two or more of the following are present: [16] (a) six or more café-au-lait spots, (b) two or more neurofibromas or one plexiform neurofibroma, (c) skinfold freckling, (d) optic glioma, (e) two or more Lisch nodules, (f) distinctive skeletal lesion, or (g) a first degree relative with neurofibromatosis type 1. Our patient had multiple neurofibroma, scoliosis, genu valgum deformity, deformities of the big toes, paraplegia, and one café-au-lait spot. Visceral involvement in disseminated neurofibromatosis is rare [9].

Involvement of the urinary tract in neurofibromatosis is rare [9]. Neurofibroma of the bladder is a rare condition with few reported cases [2,9,10,12,14,17] in the literature and neurofibromatosis of the bladder resulting in vesicovaginal fistula has not been previously reported to the best of our knowledge after a thorough literature search. The first case of bladder and spinal neurofibromatosis was reported by Gerhardt in 1878 [12]. The bladder is the commonest affected organ in the urinary tract due to the presence of a rich autonomic plexus [11] either as an isolated mass or a diffuse infiltrative process. Symptoms in urinary tract involvement are hematuria, dysuria, urinary retension and mainly irritative complaints [2,9,11]. Urinary bladder neurofibroma may present with lower urinary tract symptoms, flank pain or enuresis and incontinence [12]. This patient had urinary incontinence for which she was catheterized and later developed continuous leakage of urine despite catheterization.

Paraesthesia and paraplegia as seen in this patient has previously been described in patients with neurofibromatosis [18]. Deformity of the spine such as scoliosis and kyphoscoliosis could be found in patients with neurofibromatosis [19]. Our patient has scoliosis.

The patient was worked up for repair of vesicovaginal fistula. She was repaired using the vaginal approach. An inverted T-shaped incision was made on the fistula margins and the bladder was dissected off the anterior vaginal wall to expose the margins of the fistula. The fistula margins were closed in one layer using absorbable sutures. The anterior vaginal wall was also closed in one layer using similar sutures. Post closure dye test was negative. Postoperatively, patient was catheterized for continuous bladder drainage. She was also placed on intravenous fluids, analgesics and antibiotics. She was clinically stable and dry until the tenth postoperative day when she noticed continuous leakage of urine despite urethral catheterization. Dye test done was positive and an assessment of failed fistula repair was made. Patient was discharged on the 14th postoperative day and counselled for second repair in three months. She subsequently had a second repair after four months which was successful though patient now has stress urinary incontinence. Histology report of the fistula margins did not show bladder neurofibromatosis and also ruled out malignancy.
Consent was obtained from the patient before publication of this case report and the images presented.

Competing Interest

The authors of this publication have no competing interests.

References


