

Marjolin's Ulcer in a Spina Bifida Patient: A Case Report

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'They dress the wound of my people as though it were not serious'¹ Jeremiah 6:14.

Pressure ulcers are a frequent complication among neurologically-impaired patients, including those with spina bifida. Malignant degeneration of these pressure ulcers, known as Marjolin's ulcers, although a rare complication, results in a virulent cancer and often death. The history of a twenty year-old spina bifida patient who presented with a longstanding sacral pressure ulcer that was found to be malignant is reported. Pressure ulcers should be thoroughly investigated at presentation, to avoid labeling malignancies 'chronic ulcers', leading to delay in appropriate treatment.

Introduction

Pressure ulcers are common after spinal cord injuries (SCI). Increased motor vehicle accidents alongside improved healthcare, has led to an increase in the number of SCI survivors, and concomitantly, pressure ulcers². Although SCI and spina bifida patients are different entities, they have in common the absence of sensation and immobility, factors that place them at similar heightened risks for the development of pressure ulcers^{2,3}. As improved healthcare creeps into Sub-Saharan Africa, the number of children with spina bifida surviving and attaining adulthood is increasing. As a consequence, pressure ulcers are seen more frequently. Because pressure ulcers and ulcer recurrences in this population are such a common and difficult problems to manage², there has been a general tendency towards a degree of 'benign neglect', both by the patient and healthcare givers. This attitude is dangerous, as noted by Ratliff, who reported malignancies in the chronic pressure ulcers of two spina bifida patients referred for treatment⁴. The term 'Marjolin's ulcer' is used to describe malignant degeneration of chronic ulcers, burn scars and other chronic processes^{4,5}.

Case report

A 20 year-old African female presented with a huge foul-smelling sacral ulcer. Born with a spina bifida, she had had a bilateral below-knee amputation as a child, but was able to ambulate using bilateral prosthesis and crutches (Figure 1). Her past medical history was sketchy, and with no medical records available, was reconstructed from memory. Except for occasional visits to dispensaries and hospitals, home wound care consisted only of pieces of clothing used to cover the ulcer and absorb its exudate. She had urinary bladder and bowel incontinence. She developed a sacral pressure ulcer at the age of four. The ulcer smoldered over the subsequent 16 years, gradually growing bigger, unable to access appropriate care. Because she had no perineal sensation, she had no pain from the ulcer, but was aware of the foul smell emanating from the ulcer.

She was referred to our institution by a 'Good Samaritan', who also paid for her treatment. On examination, the ulcer edges were indurated, elevated, but inverted. The ulcer base and walls had multiple elevated lesions, extending over a large undermined area, with multiple sinuses (Figure 2). The perineum was macerated and patched with areas of vitiligo by urine. She had no palpable nodes. An initial biopsy of the pressure ulcer revealed a squamous cell carcinoma (Marjolin's ulcer). A pelvic/abdominal ultrasound and a chest radiograph did not show any evidence of metastases. A wide excision and wound closure using local flaps were performed. The histopathology reported a squamous cell carcinoma deeply invasive, extending into bone, but with clear margins. Wound infection and dehiscence in the first week post-operatively was debrided and closed primarily.



Figure 1. Bilateral amputatim.



Figure 2. Sacral pressure ulcer. Note multiple fistuli with purulent discharge – all communicated with the ulcer. Note also chronic skin changes, including perineal vitiligo from repeated exposure to urine.

The wound healed completely over the ensuing four weeks, with no evidence of local tumor recurrence at 5 months. She received no further treatment, but went back to normal life, rid of the foul smell, and with enhanced social relations. Prior to her surgery, the patient had been secluded from social interaction, preferring to stay indoors because of the odour emanating from her ulcer.

Post-operatively, for most of the ten months that she lived post-operatively, she ran a small business: this period that may have been the best time of her short life. No additional tests were done after her surgery, because of cost constraints. She died at home, and though no autopsy was performed, death due to metastatic disease was presumed, based on the history given by relatives on her last few weeks of her life.

Discussion

Malignant degeneration of chronic ulcers, usually into squamous cell carcinomas was first described by Jean Nicolas Marjolin in 1828. Marjolin's ulcers have since been reported in burn scars, chronic osteomyelitis, post-traumatic wounds and chronic fistuli^{5,6}.

The causes of malignant degeneration are not known, but a few theories have been proposed. The initiation and promotion theory proposes a process of transformation of normal cells into dormant malignant cells, with subsequent cellular promotion and tumor growth, with infection acting as a co-carcinogen. The chronic irritation theory suggests malignant transformation results from cycles of repeated irritation, trauma and attempted repair. Toxins released by chronic ulcer cells may act as carcinogens, leading to development of tumors. Traumatic implantation of epidermal cells into the dermis resulting in foreign body reaction and ultimately malignant transformation is another hypothesis. The relatively avascular area of scars may interfere with immune surveillance, leading to uncontrolled proliferation of immunologically undetected tumor cells⁵⁻⁸. Spina bifida, with the associated lack of sensation and immobility are congenital in origin, while SCIs are acquired, many in

young adulthood. Although the proposed theories are applicable to ulcers in both SCI and spina bifida patients, it is feasible that different mechanisms may be at play in pressure ulcers of these two populations, resulting in the differences in rates of malignant degeneration.

While squamous cell carcinoma is the most commonly observed Marjolin's ulcer type, basal cell carcinoma, adenocarcinoma, sarcomas, melanoma and verrucous carcinoma have also been reported. Malignant degeneration of pressure ulcers though rare, is well described⁴⁻¹⁰. Mustoe found a 0.5% incidence of Marjolin's ulcer amongst patients with pressure ulcers⁵. Most Marjolin's ulcers occur in the sacral and ischial areas. Although malignant degeneration in a pressure ulcer has been reported to have occurred after 6 months, the average latency period of about 20 years is much shorter than that in burn scars (31 years)^{5,6}.

Malignant transformation of pressure ulcers frequently leads to the death of the patients. These sadly, are preventable deaths – either by the prevention of pressure ulcers, or early and effective management, should they occur^{2,4,5}. Some workers have suggested that these are immunologically privileged tumors that overwhelm the patient's immune system upon surgical manipulation, leading to systemic metastasis and death⁵, as may have been the case with our patient. Marjolin's ulcers in burn scars or chronic osteomyelitis, with no evidence of metastases have a much better prognosis than those in pressure ulcers⁷. Although tumor-negative margins are generally reassuring in surgical oncology, this may not be true for the virulent pressure ulcer carcinomas⁸. Wide excision with elective nodal dissection, or even hemi corporectomy as indicated, have been proposed for pressure ulcer carcinomas, when cure is intended^{8,9}. Radiotherapy and chemotherapy, although frequently used post-operatively, have not been shown to be effective^{5,8}.

This is the first report of Marjolin's ulcer in a spina bifida patient from Africa. Although pressure ulcers are common amongst spina bifida patients, the incidence of Marjolin's ulcers in this population is extremely rare. A search of English literature revealed a total of six reported cases of Marjolin's ulcers, in four articles^{4,8-10}. This rarity is difficult to explain. While Marjolin's ulcers generally affect patients with poor access to healthcare, all previous reports were from developed countries, indicating that vigilance is required, irrespective of the economic environment. All pressure ulcers should be thoroughly investigated at presentation, to avoid labeling malignancies 'chronic ulcers', leading to delay in appropriate treatment.

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Female Urethral Leiomyoma: A Case Presentation

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We present a case of female urethral Leiomyoma. The mass which protruding from the urethral meatus causing dysuria and urethrorragia. Our physical examination revealed the presence of the mass on the anterior wall of the vagina protruding from the urethral meatus. Histopathological examination showed leiomyoma of the female urethra. The patient was treated surgically and the symptoms disappeared completely.

Introduction

Leiomyomas are benign tumors of smooth muscle origin and rarely found in the urinary tract, with only 40 cases of urethral leiomyoma reported in the literature^{1,2,3,4,9}. It is more common in women between 30 and 50 years old^{1,3,5}. The first case was reported in 1984 by Buttner⁶. There is a controversy about its dependency on estrogen hormones. Its diagnosis is made only by histopathologic examination. The topographic site of the lesion is very useful for the prognosis. Grabstald and Cols⁷ classified the tumors as distal or anterior when it's located at the distal 1/3 of the urethra, and proximal or posterior when it's located at anterior 1/3. We report a case of leiomyoma of the female urethra with its clinicopathological characteristics.

Case Report

A 27 years old female patient with no history of past illness, came to our clinic presenting a mass protruding from the urethral meatus for 18 months, accompanied with dysuria and urethrorragia. On physical examination there was a polyploid and rounded mass with the size of 3.5 cm in diameter, smooth surfaced, firm with pink colour, located at the distal posterior urethra. On straining it protrudes through the urethral meatus (Figures 1). All investigations were normals.

FNA Biopsy done revealed Epidermid cyst of the urethra. She was operated, tumour excised and specimen sent for histopathologic examination (Figures 2).



Figure 1. Leiomyoma of Female Urethra Before Surgery



Figure 2. Macroscopic View of the Tumour

The histopathologic examination showed a leiomyoma of the urethra. On follow after surgery the patient was found to be asymptomatic with out any abnormal finding on urologic examination.

Discussion

Leiomyoma is a benign tumor of mesenchymal origin, composed of smooth muscle cells found rarely in the urinary tract. Our patient histopathologic result is also same. It is known to involve in decreasing order of frequency, the kidney bladder and urethra^{3,13,14}. Leiomyomas are 3 times more common in women between 30 and 50 years^{1,3,5}. According to the mentioned criteria, the case presented was atypical because it appeared in a 27 years old lady. The diameter of the tumors ranged from 1 – 40 cm^{2,3,14,15}. The pathogenesis of leiomyoma is unknown but its growth is probably endocrine dependent, with the growth patterns and size influenced by estrogen^{2,3,8,15}. But our case did not have hormone dependency because as we mentioned above that she is young with no such abnormality on investigations and there is no history of oestrogen based contraceptives usage. The clinical presentations depends on the location and size of the tumor. Patient can be asymptomatic being the leiomyoma an incidental finding during gynecological examination. Common presenting symptoms are periurethral masses, urinary tract infection, hematuria, urethrrragia, dysuria and dyspareunia and even acute urine retention and acute renal failure^{2,3,4,10,12} patient also had two of the mentioned clinical presentation.

The clinical diagnosis is made by history, physical examination, uretherocystoscopic examination and imaging studies like transvaginal sonography, retrograde urethrography, voiding cystourethrogram and MRI^{2,3,4,10,12,13,14}. The histopathologic study will give the definitive diagnosis. We also reached on definitive diagnosis by histopathologic examination. The differential diagnosis of female urethral leiomyoma should be done with urethral caruncle, papilloma, urethral diverticulum, ectopic ureterocele, fibrous polyp, Grätner's duct cyst, periurethral abscess, urethral carcinoma and other mesenchymal tumors^{3,14}.

The urethral leiomyoma is treated surgically without recurrence^{1,3}. Even we treated the patient surgically and she is asymptomatic with no recurrence. The operation techniques depends on the site of the tumor. So we removed the masas through incision of the anterior vaginal wall. The prognosis of this tumor is excellent as malignant transformation has not been reported^{1,2,3,8}. Our patient is also doing good with no complication or recurrence after surgical therapy.

Conclusion

1. Leiomyoma of the female urethra is a benign mesenchymal tumor and rarely found in the urinary tract.
2. The diagnosis is always confirmed by histopathological study.
3. The treatement is always surgical.

4. The prognosis is excellent since it has no risk of malignant transformation.

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