Isolated Giant Vulval Neurofibroma: A Case Report

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Abstract

A case of isolated giant vulval neurofibroma in a 34 year old para 1 lady is reported. She presented with a large vulval swelling involving labia majora, minora and the clitoris preventing her from having sexual intercourse. She had vulval biopsy sent for histopathological examination and it was diagnosed as "chronically inflammed neurofibroma of the vulval". Because of the large size, simple vulvectomy was performed and she recovered well postoperatively. Literature on vulval neurofibroma regarding the presentation and management was revieved.

Key Words: Vulva Neurofibroma, Vulvectomy [Trop J Obstet Gynaecol, 2004;21:188-189]

Introduction

Neurofibromas (schwannomas) are generally small, soft polypoidal lesions occurring in children and young adults, and if present in the vulva, it represents a local manifestation of von Recklinghausen's disease in up to half of cases[1]. They arise from the neural sheath and are usually of no consequences but when they are multiple could disfigure the vulva and interfere with sexual function[2]. Isolated vulva neurofibroma is an uncommon tumour and only few large ones had been reported. This case of giant localised vulval neurofibroma managed at Olabisi Onabanjo University Teaching Hospital, Sagamu Nigeria is reported.

Case Report

Mrs. A.O. a 34 year-old para 1 + 1 lady presented at our gynaecological out patient clinic about 4 years ago with a vulva swelling of six month duration. It was small and without any ulceration. In the course of investigating her, she defaulted and did not attend the clinic again until four years later. At the time, the mass was much larger with extensive ulceration over its surface. It was making her walking uncomfortable and had prevented her from having sexual intercourse since the last three years. During the period she defaulted, she attended various traditional medical places for various treatments, all to no avail. It was when the mass was getting bigger despite the traditional treatment and she also developed ulceration with offensive odour that she decided to come back to the hospital.

She had only a child delivered vaginally about eight years ago and a spontaneous abortion of a ten week old pregnancy about years after delivery. Her menarche was at 16 years and had a moderate flow for four days in a regular cycle of 27 days. She had no significant medical history and had not been operated on in the past. Her family history was not remarkable for such or other swellings in any part of her body, and in her social history, she neither drinks alcohol nor smoke tobacco.

On examination, her general condition was satisfactory. She was not pale and afebrile. Her blood pressure was 100/60mm Hg and her pulse rate was 72 beats per minute, regular and of good volume. Her lung fields were clinically clear and heart sounds I and II were heard with no murmurs. Her abdomen was normal. She had no inguinal lymphadenopathy. She had no 'cafe au leit' spots. The significant findings were in the vulva. The vulval was grossly enlarged and the enlargement involved both labia majora and minora, but the left was about twice the size of the right; 16 inches by 10 inches and 9 inches by 7 inches respectively. Some areas on the surfaces were sclerotic while others were hyperkeratotic. There were multiple decubitus ulcers in the dependent parts of the masses. See Picture 1. The vaginal orifice was blocked by the masses and entry could only be gained into it by displacing the masses laterally.

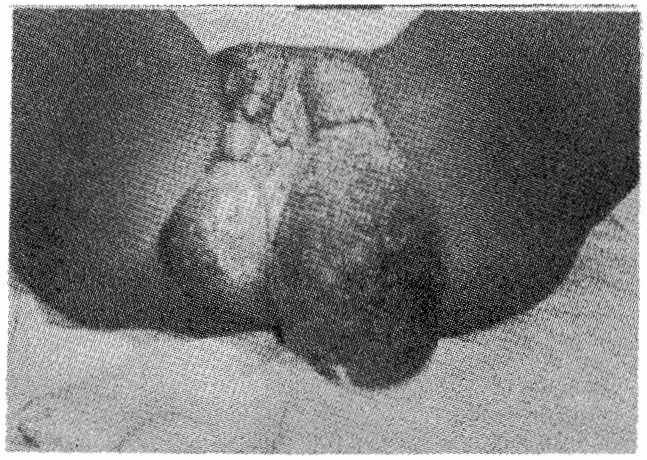

Fig. 1

The cervix was however normal, the uterus was normal, the adnexa and pouch of Douglass were normal. The findings were explained to her and the following investigations with their results were carried out.

Full blood count

Packed cell volume was 32%
Total white cell count was 5500/mm^3

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Different white cell count		
Neutrophils		62%
Lymphocytes		37%
Eosinophils		1%
Blood for microfilaria	-	Negative
Skin snip for microfilaria	-	Negative

Urinalysis was normal

Chest Xray. Electrolytes, Urea and Creatinine were normal; and she was negative for both HIV I and II. She had biopsy of the vulva mass with local lignocaine (2%) infiltration. The biopsy specimen was sent for histopathological examination. The report of the biopsy was 'chronically inflammed neurofibroma of the vulval'.The need for surgical removal and the extent of the operation were explained to her and she consented to the operation. Because of the size of the mass preventing her from having sexual intercourse, the chronicity and involvement of both labia majora, minora and clitoris, a simple vulvectomy was carried out. Refer to Picture 2. The mass weighed 1500g (1.5 kilogrammes).

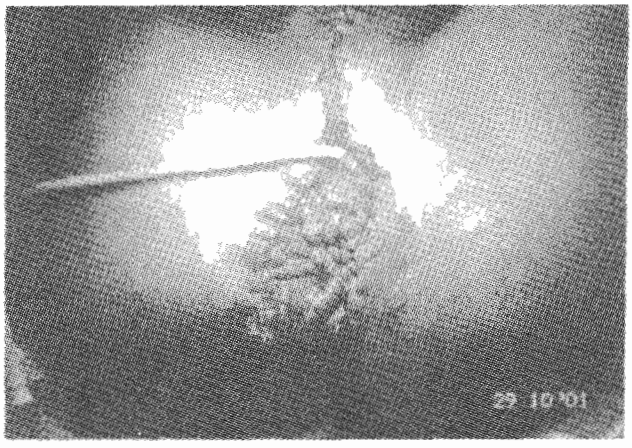

Fig. 2

A few polypoidal growths protruding from the anus were found. She recovered well post-operatively and was discharged home on the fourteenth postoperative day. The histopathological report of the mass confirmed neurofibroma of the vulva. She was seen at the gynaecological out patient clinic four weeks after discharge. She had no complaint, the vaginal orifice was now obvious and the wound had healed well. She was asked to see the general surgical team for assessment of the anal polypoidal growth.

Discussion

Neurofibroma of the vulva alone is unusual and only few of such cases had been reported in literature [3-7]. It usually occurs as part of manifestations of von Recklinghausen's disease with variable involvement of the labia majora, minora and clitoris. The case reported occurred without any of the features of von Recklinghausen's disease. Unlike neuroma of the vulval after episiotomy which is postulated to be as a result of disorganised proliferation of proximal nerve stumps following episiotomy[8], the aetiology or predisposing factors in vulva neurofibroma apart from being part of manifestation of von Recklinghausen's disease are not known. In the patient presented, there was neither identifiable aetiological or predisposing factor nor was it part of von Recklinghausen's disease, as no other feature both cutaneous or otherwise was found in her.

Though neurofibromas are usually small in size[1], the case presented attained a big size of 1500 grammes. This could have been due to continuous growth over a long period of more than four years when she defaulted from coming to hospital. The big size of the mass between the thighs with accompanying friction while she walked about and the dependency of the tip predisposed it to ischaemic necrosis, producing ulcers on its surface which subsequently became infected. Though excision is usually carried out for vulva neurofibroma, when the tumour is very large and interferes with sexual function, it may require vulvectomy[2], as was done for this case because of its big size. She recovered well portoperative and the vagina was now accessible for sexual intercourse.

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