Plexiform Neurofibroma of the Penis and Facial Nerve

L Lawson Douglas, CAM Cadogan

ABSTRACT

A case of plexiform neurofibroma of the penis is presented. It is a rare condition often found in association with congenital neurofibromatosis. This case is unique because of the accompanying lesion of the facial nerve above the right eye, an association not previously reported. The patient was managed effectively by adequate resection of the penile lesion.

Keywords: Penis, plexiform neurofibroma

Neurofibroma Plexiforme del Pene y del Nervio Facial

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RESUMEN

Se presenta un caso de neurofibroma plexiforme del pene. Se trata de una condición rara, a menudo asociada con una neurofibromatosis congénita. Este caso es único debido a la lesión acompañante del nervio facial sobre el ojo derecho – una asociación no reportada con anterioridad. El paciente fue tratado con efectividad mediante la resección adecuada de la lesión del pene.

Palabras claves: Pene, neurofibroma plexiforme

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INTRODUCTION

Plexiform neurofibroma (PN) is a variant of neurofibroma that presents as a swelling of the peripheral nerves and may become massive in size. It is a congenital lesion and tends to occur in the orbit, neck, back and inguinal region. Lesions involving the penis are uncommon (1, 2). We report a case of PN of the penis occurring in association with a similar lesion of a branch of the facial nerve.

CASE REPORT

An eight-year old boy was seen in the urology clinic at the Kingston Public Hospital for evaluation of a swelling of the penis. It had increased in size over a few years. He was otherwise healthy.

On examination, there was a large soft tissue mass that arose symmetrically from the mid-shaft and extended distally to involve the corona. The glans was normal (Figs. 1, 2).

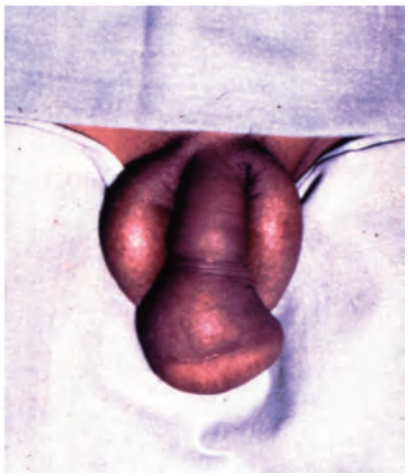

Fig. 1: Dorsal view of the penis.

Also of note was a one centimeter swelling in the upper outer quadrant of the right orbit (Figs. 3, 4) that had a similar history of growth to the penile lesion. Biopsies of both lesions were reported as PN. The patient was referred to plastic and reconstructive surgery and the penile lesion was successfully excised with a good cosmetic result.

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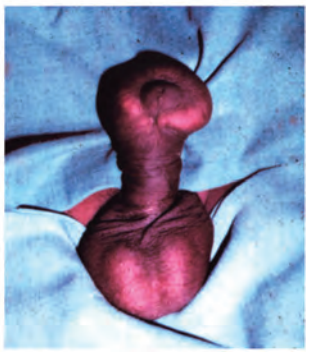

Fig. 2: Ventral view of the penis.

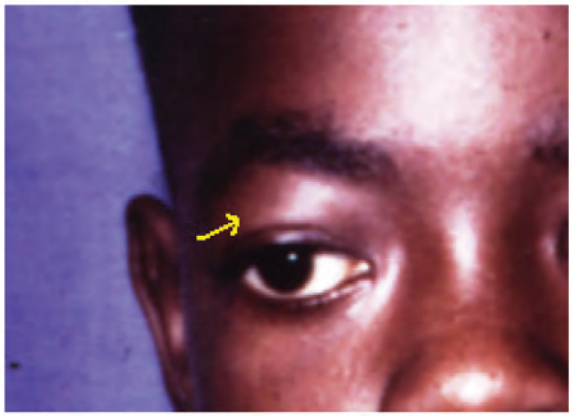

Fig. 3: Anterior view of the right eye.

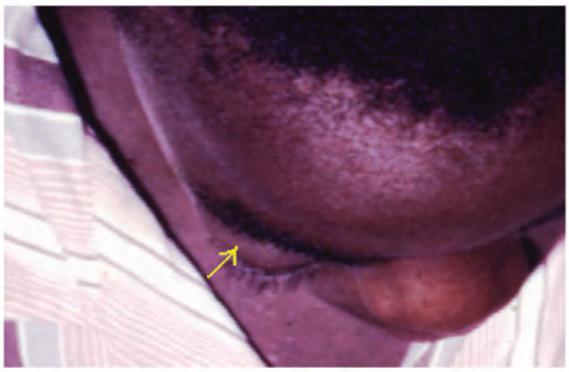

Fig. 4: Superior view of the right eye.

DISCUSSION

Plexiform neurofibroma is a variation of neurofibromatosis and is classified as a phakomatosis belonging to the group of neurocutaneous syndromes which also include von Recklinghausen's disease, Hippel Lindau disease, neurocutaneous melanosis and Sturge Weber syndrome. These diseases may be closely related and sporadic examples of patients with more than one of these have been reported (1, 2).

Plexiform neurofibroma of the penis in children is rare and in 2007, only eight cases had been reported (1, 3, 5−7). Some patients had café-au-lait spots but in others the penile lesion was the only manifestation of the disease (5, 6). Plexiform neurofibroma may undergo malignant change but the risk is thought to be only 5−16%. The aim of treatment should be complete excision and ongoing follow-up. Periodic physical examination and magnetic resonance imaging are recommended (5).

In some cases, the lesions were small and easily excised but others required extensive excision (often incomplete) or partial penectomy because of size and involvement of the tunica (4, 6, 7). This appears to be the only reported case occurring in association with a lesion of the facial nerve. The patient was referred to plastic and reconstructive surgery in an effort to achieve the best cosmetic result. The functional result was not determined.

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