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CASE REPORT

Clinical case: Von Hippel-Lindau disease, a nursing perspective☆

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KEYWORDS
Nursing care;
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Abstract
Introduction: Von Hippel-Lindau disease (VHL) is an autosomal dominant disease caused by a mutation of the VHL tumour suppressor gene. Patients with VHL may have cerebellar haemangioblastomas, retinal haemangioblastomas, phaeochromocytomas, renal carcinoma, pancreatic cysts, and pancreatic neuroendocrine tumours. The aim of this case report is to adapt the plan of nursing care to the patient's needs.
Method: This is a clinical case of a patient admitted to the neuro-rehabilitation unit. The clinical history was analysed by collecting the demographic/clinical data with prior consent.
Results: A male patient (49 years) was admitted to a neuro-rehabilitation unit for therapy after surgical excision of a haemangioblastoma of the third ventricle, as a result of VHL disease. A thorough care plan tailored to the patient's individual needs was established using NANDA-NIC-NOC nursing taxonomy. The following nursing diagnoses were identified: unilateral neglect (0123), poor knowledge (0126), acute confusion (0128), impaired verbal communication (0051), ineffective control of impulses (0222), self-care deficit: bathroom (0108), clothing (0109), and use the toilet (0110), inefficient management of health (0078) and deterioration in walking (088). At discharge, the diagnosis criteria were developed positively, and the results were improved.

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Conclusion: Due to VHL syndrome being considered a rare disease, knowledge of the pathophysiology has allowed us to develop a care plan that identifies the health problems in order to provide adequate nursing care to patients and their needs.

PALABRAS CLAVE
Atención de Enfermería;
Diagnósticos enfermeros;
Planificación de atención al paciente

Caso clínico: síndrome de Von Hippel-Lindau, una visión desde enfermería

Resumen
Introducción: La enfermedad de Von Hippel-Lindau (VHL), es una mutación heredada del gen de supresión tumoral pVHL. Los pacientes con VHL pueden presentar hemangioblastomas cerebelosos, hemangioblastomas retinianos, feocromocitomas, cáncer de riñón, quistes pancreáticos y tumores neuroendocrinos pancreáticos. El objetivo del presente caso clínico es adaptar el plan de cuidados de enfermería a las necesidades presentadas por el paciente.
Método: Se elabora el caso clínico de un paciente ingresado en la unidad de neurorrehabilitación. Se analizó su historia clínica para la recogida de datos demográficos/clínicos con su consentimiento previo.
Resultados: Se trata de un varón de 49 años que ingresa en la unidad para tratamiento neurorrehabilitador por intervención quirúrgica de hemangioblastoma de suelo del tercer ventrículo a causa del síndrome de VHL. Se ha efectuado un plan de cuidados adaptado a sus necesidades con la taxonomía enfermera de la NANDA-NIC-NOC. Se identificaron los siguientes diagnósticos de enfermería (DdE): desatención unilateral (0123), conocimientos deficientes (0126), confusión crónica (0129), deterioro de la comunicación verbal (0051), control de impulsos ineficaz (0222), déficit de autocuidado: baño (0108), vestido (0109) y uso del inodoro (0110), gestión ineficaz de la propia salud (0078) y deterioro de la ambulación (088). Al alta, los criterios de resultado de gran parte de los DdE activados al ingreso evolucionaron positivamente.
Conclusión: Dado que el síndrome de VHL se considera una enfermedad rara, el conocimiento del presente caso ha permitido elaborar un plan de cuidados que permite identificar los problemas de salud para proporcionar unos cuidados de enfermería adecuados al paciente.

Introduction

Von Hippel-Lindau disease (VHL) is a hereditary, autosomal dominant disease caused by a mutation in the tumour suppressor gene pVHL, whose function is to regulate the vascular endothelial growth factor.[1–3] Loss of this function results in a series of tumour malformations; notably haemangioblastoma of the central nervous system, capillary haemangioma of the retina and clear cell renal carcinoma.[1,3] In Europe, the prevalence of VHL syndrome varies between 1/39,000 and 1/53,000 with an estimated incidence of 1/36,000 live births.[1,2] Haemangioblastomas of the central nervous system are the most common tumours in VHL syndrome and affect between 60% and 80% of patients diagnosed with the disease between the ages of 40 and 50 years. Despite the benign nature of cerebellar haemangioblastoma, it can progress to major neurological and functional disorders.[2,4] Its complete surgical resection is usually feasible, however, the neurological sequelae as a result both of the progression of the disease and/or surgical intervention, make patients with VHL syndrome candidates for neurofunctional rehabilitation in order to minimise these sequelae.[4]

The objective of this clinical case is a description of the nursing care plan tailored to the needs of an individual patient with a rare disease.

Development of the clinical case

A patient was selected who had been admitted to the neurorehabilitation unit of a Madrid hospital in 2015. The clinical and demographic data were collected retrospectively from his clinical history after his prior consent.

Description of the case

A 49-year-old male, referred to the neurorehabilitation unit for rehabilitation treatment necessary after surgical intervention for a haemangioblastoma of the floor of the third ventricle caused by VHL syndrome. The patient had a history of arterial hypertension, was an ex-smoker, had undergone bilateral nephrectomy for clear cell renal carcinoma (treated with haemodialysis), and presented anaemia, nodular lesions in the hypothalamus (growing) and IV