CASE REPORT

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

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KEYWORDS
Nursing care;
Nursing diagnoses;
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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.
Clinical case: Von Hippel-Lindau disease, a nursing perspective

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.
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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 neuorrehabilitación. 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 neuorrehabilitador 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 (DoE): 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 DoE 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 un cuidados de enfermería adecuados al paciente.
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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 neuorrehabilitation 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 neuorrehabilitation 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...
ventricle (stable). No known drug allergies. The patient's vital signs on admission were within normal limits: blood pressure, 100/60 mmHg, cardiac frequency, 64 lx, basal oxygen saturation, 97%.

A nursing assessment was carried out on admission, based on the Virginia Henderson model, to establish the patient's needs:

- Oxygenation:
  - Eupneic, no alteration in respiratory pattern, oxygen therapy not required.
- Nutritional/hydration:
  - Swallowing and chewing unimpaired.
  - Low potassium diet for dialysis patients
  - Partially dependent for this activity.
- Elimination:
  - Anephric.
  - Continent of faeces and incontinence pads not required.
- Movement and posture:
  - Confined to bed or chair.
  - Able to collaborate with transfers.
  - Barthel's index: 30 "severe dependence".
- Sleep/rest:
  - No alteration in this pattern.
  - Medication to help with sleep not required.
- Dressing/undressing:
  - Partially dependent for this activity.
- Heat regulation:
  - No heat regulation disorder.
- Hygiene:
  - Dependent for this activity.
  - Arteriovenous fistula for haemodialysis.
- Safety:
  - No pressure ulcers. According to Braden–Bergstrom Scale (score 13), "moderate risk for developing a pressure ulcer".
  - Mechanical securing in chair required (abdominal fastening belt), due to risk of falling. Downton Scale "high risk".
- Communication:
  - Conscious and alert on admission, Glasgow Coma Scale: 12.
  - Pfeiffer Scale "moderate intellectual disability".
  - Presenting nomination, comprehension and repetition, and bradylalia and bradypsychia.
- Beliefs and values:
  - No manifestation of religious or spiritual beliefs.
  - Married with two daughters, also diagnosed with VHL.
- Work/fulfilment:
  - Employed in the building industry.
- Leisure/learning:
  - Leisure activities are watching television and listening to the radio.

Results

An initial nursing care plan was drawn up (Table 1) by means of the initial patient assessment and anamnesis based on the problems identified using the NANDA, NIC and NOC taxonomies. The following nursing diagnoses (ND) were made on admission: unilateral neglect (0123), deficient knowledge (0126), acute confusion (0128), impaired verbal communication (0051), ineffective control (0222), self-care deficit: bathing (0108), dressing (0109) and toileting (0110), inefficient management of own health (0078) and impaired walking (088). The nursing care plan was adapted to the patient's needs as they arose through continuous evaluation based on his progress.

The patient was discharged home with his wife after 6 months' hospitalisation and neurorehabilitation treatment managed by an interdisciplinary team (nurse, care assistant, doctor, physiotherapist, occupational therapist, neuropsychologist, speech therapist).

On discharge, the outcome criteria of most of the nursing diagnoses that were activated on admission had evolved positively. With regard to ND (088) impaired walking, the patient managed to perform transfers alone (NOC [208] mobility, indicator [30101]), and was able to stand and walk with no technical aids, under supervision (indicator [20806] walking, Likert 1 > Likert 4). The nursing diagnoses: self-care deficit: bathing (0108), dressing (0109) and toileting (0110) also had a positive outcome, needs personal help to dress (NOC [0302] self-care: dressing, indicators: [30204] the patient managed to dress the upper part of his body and [30205] dress the lower part of his body, Likert 1 > Likert 4), for bathing (NOC [0301] self-care: bathing, indicator [30109] washed in the shower, Likert 1 > Likert 3) and used the toilet (NOC [0310] self-care: toileting, indicator [31013] entered and left the bathroom, Likert 1 > Likert 4).

In terms of language, the patient presented nomination, comprehension and repetition, with mild bradylalia associated with bradypsychia (NOC [0902] communication, indicator [90202] used spoken language, Likert 1 > Likert 4). In relation to the ND of chronic confusion (0129), at time of discharge the patient continued to have mild/moderate cognitive impairment which had improved in only a few indicators (NOC [0901] cognitive orientation, indicators: [90101] he was able to identify himself, Likert 1 > Likert 5; [90103] identify place, Likert 1 > Likert 3; [90104] and identify time Likert 1 > Likert 3) (Table 2).

Discussion/implications for clinical practice

VHL is a congenital condition that causes malignant tumours in multiple organs, such as cerebellar haemangioblastoma. Due to the gradual progression of the disease leading to neurological and functional disorders, nursing care, psychological support and the individualised application of a neurorehabilitation programme (physiotherapy, occupational therapy, treatment from the neuropsychologist, speech therapy) proved majorly important in improving this patient's quality of life. Therefore, and given that these tumours caused by VHL are extremely rare and of uncertain prognosis, we stress the essential and priority role of nursing action in an integral approach to these patients and their families.

Standardising a care plan to encompass the condition's neurological damage in all of its phases helps greatly towards standardising language and nursing records. Since the proposed interventions are based on better knowledge and experience on the part of nursing staff, providing
<table>
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<th>NANDA</th>
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<th>Activities</th>
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<tr>
<td>00126: deficient knowledge related with impaired cognitive function triggered by insufficient knowledge</td>
<td>1811: knowledge: prescribed activity. 1823: knowledge health promotion</td>
<td>5606: teaching: individual</td>
<td>560603: to establish the patient’s teaching needs. 560610: set mutual, realistic learning goals with the patient. 560614: select appropriate teaching methods/strategies for the patient. 560619: instruct the patient when appropriate</td>
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<td>00129: chronic confusion related with brain lesion triggered by progressive impairment of cognitive function</td>
<td>1214: level of agitation 901: cognitive orientation</td>
<td>6486: environmental management: safety 6490: fall prevention 4720: cognitive stimulation</td>
<td>648601: identify safety needs, based on the patient’s physical and cognitive function and behavioural history. 648603: eliminate environmental risk factors, when possible. 649015: immobilise wheels of wheelchair, beds or stretchers when transferring the patient. 649025: use guard rails of an appropriate length and height to prevent falls from the bed, if necessary. 472007: give guidance as to time, place and person. 472012: place familiar objects and photographs in the patient’s environment 492002: show interest in the patient. 492005: show emotional awareness and sensitivity. 492017: use silence/listening to encourage the patient to express their feelings, thoughts and concerns. 497608: adapt communication style to meet the patient’s needs (e.g., be in front of the patient when talking to them, listen carefully, present an idea or thought each time, speak slowly and avoid shouting, use written communication or ask for assistance from the family to understand the patient’s speech). 497615: collaborate with the family and speech therapist to develop a plan to enhance effective communication 435006: establish habits. 435007: ensure consistency between shifts with regard to care environment and routine. 435021: apply wrist/leg/chest bands, if needed</td>
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<td>0051: impaired verbal communication related with bradypsychia triggered by abnormally slow in language and word articulation</td>
<td>902: communication</td>
<td>4920: active listening 4976: communication enhancement: speech deficit</td>
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<td>00222: ineffective control related with impaired cognitive function triggered by acting without thought</td>
<td>1405: impulse self-control</td>
<td>4350: behaviour management</td>
<td>435006: establish habits. 435007: ensure consistency between shifts with regard to care environment and routine. 435021: apply wrist/leg/chest bands, if needed</td>
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<td>00108: self-care deficit: bathing related with neuromuscular impairment triggered by impaired ability to wash body</td>
<td>301: self-care bathing 305: self-care: hygiene</td>
<td>1801: self-care assistance: bathing and hygiene 1610: bathing</td>
<td>180103: provide support with tooth brushing, if necessary. 180104: enable the patient to wash independently, as appropriate. 180108: provide help until the patient is fully able to self care 161001: help with showering on chair, bathtub, bed bath, standing shower or sitting bath, as appropriate or desired. 161012: check skin condition during bathing</td>
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<tr>
<td>00109: self-care deficit: dressing related with neuromuscular impairment triggered by impaired ability to put on and take off different items of clothing</td>
<td>302: self-care: dressing</td>
<td>1630: dressing 1802: self-care assistance: dressing/grooming</td>
<td>163001: identify the areas where the patient needs help with dressing. 163002: observe the patient’s ability to dress. 163012: provide assistance until the patient can take full responsibility and dress independently. 180204: be available to help with dressing, if necessary. 180205: help the patient to brush their hair, if necessary. 180206: enable the patient to shave, as appropriate. 180208: help with shoelaces, buttons and zips, if necessary</td>
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evidence-based care will help to reduce variability in care practice.

Conclusion

The neurorehabilitation programme applied to the patient with VHL disease enabled him to partially recover his motor functions. The patient was able to carry out basic activities of daily living with minimal personal help. Similarly, knowledge of the pathophysiology and progressive evolution of VHL syndrome, a disease that is considered rare, enabled us to prepare a nursing care plan tailored to the problems that we identified using the NANDA, NIC and NOC taxonomies, and to individualise nursing care to achieve measurable results.
and provide continuous assessment of the outcomes of the initially activated nursing diagnoses.

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References