Micrograph of a schwannoma, which is a tumour seen in neurofibromatosis type 2

Neurofibromatosis type 2 (NF2) is a rare genetic condition with a birth incidence of one in 35,000 (Neuro Foundation, 2013). It is an autosomal dominant genetic condition so only one copy of the gene needs to be inherited from a parent for a child to be affected. NF2 can also occur in a family with no previous history of the condition and around half the people who have it will be the first in their family to be affected. In these cases, the cause is a misprint on chromosome 22 at conception.

**Diagnosis**

Bilateral vestibular schwannomas – benign tumours on the hearing and balance nerves – are major features of NF2. The early signs are hearing loss, often in the late teens or early 20s. About 60% of patients present with unilateral hearing loss. At the time of onset, bilateral vestibular schwannomas may also cause tinnitus, dizziness and poor balance (Evans et al, 1992). Ophthalmic abnormalities are also very common (Parry et al, 1994) and around 70% of patients also have skin tumours. These plaque-like lesions are slightly raised and more pigmented than the surrounding skin, often with excess hair.

Further information about diagnosis can be found at The Neuro Foundation’s website (Box 1).

**Management**

Patients should be managed in specialty centres (Evans et al, 2005). Many specialties are involved in patient care including: neurosurgery; ear, nose and throat; audiology; neurology; ophthalmology; and genetics.

All individuals who are affected should be offered a genetic test. This has implications for other family members, who may be at risk of inheriting the condition.

Patients benefit from rehabilitation courses to help reduce feelings of isolation and negotiate the practical and emotional challenges associated with hearing loss.

**Implications for general nurses**

What should nurses in A&E or on wards consider when a patient with NF2 is admitted? What are the implications for their care? Most importantly, what would the patients with NF2 most like nurses to understand about them?

Good communication is essential. Take time to find out if they are hearing impaired, for example, is one ear affected or are they completely deaf? Check whether there are any visual impairments. Ensure all members of staff are aware of any issues regarding deafness, visual impairment and poor balance. These can be frightening to experience, and particularly so when in unfamiliar surroundings.

Many patients develop a facial palsy, which can make eating difficult and embarrassing if food or saliva escapes while they are eating. Some manage better if food is minced and this should be discussed with the patient.

Eye involvement can mean that eyelids do not close fully, which can lead to ulceration and permanent damage to the cornea. Artificial tears and lubricating creams are vital to keep the eye moist and healthy.

Any concerns or queries about a patient should be directed to the local team or specialist nurses involved in the patient’s care. Patient notes should indicate who this is and patients will usually have contact numbers for their NF2 nurse. NT