

**One sentence:** The accessory nerve, or CN XI, is a motor nerve that stems from the brainstem and cervical cord, and it is involved in complex eye tracking movements.

Genetic testing	NCV/EMG	Laboratory	Imaging	Biopsy
	+		+	

## Symptoms

Weakness of the shoulder and shoulder drop. Damage to the accessory nerve can cause shoulder pain of variable severity over the shoulder and scapula.

## Signs

Trapezius muscle weakness causes shoulder drop, atrophy of trapezius muscle. Inability to lift the shoulder and raise the arm above the horizon-

tal plane. If affected, atrophy and weakness of the sternocleidomastoid muscle and impaired head rotation to the opposite side. Scapular winging (medial margin).

## Specific Qualities

*Motor:* Most of the motor supply to the trapezius muscle is derived from the accessory nerve, with contribution from the cervical plexus [1].

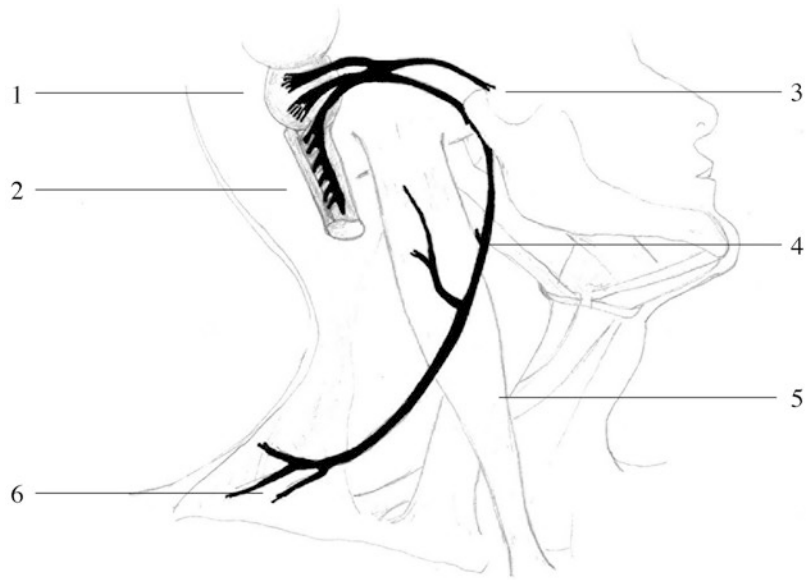
*Sensory:*

*Autonomic:*

*Special senses:*

*Other:* The accessory nerve is the only cranial nerve which enters and exits the skull. Anatomically, a distinction between the brainstem and spinal fibers is made. The “transitional nerve” is involved in laryngopharyngeal innervation. The sternocleidomastoid muscles have a prominent role in oculomotor tracking (Fig. 16.1).

**Fig. 16.1** Accessory nerve. 1 cranial roots, 2 spinal roots, 3 branch to soft palate, 4 accessory nerve, 5 sternocleidomastoid muscle, 6 trapezius muscle



## Location of Lesion

*Central:* The “central” unilateral supranuclear lesions tend to cause mild and transient weakness, as the accessory nerve nuclei receive bilateral cortical input. Hemispheric lesions rarely cause a clinically relevant CN XI paresis.

“Dissociated weakness” of the sternocleidomastoid and trapezius muscles have been reported in brainstem lesions.

*Intracranial within the skull:* Infections; tumors, e.g., schwannoma.

*Exit of the skull:* At the jugular foramen: Lesions occur in association with the glossopharyngeal and vagus nerves, e.g., Vernet’s syndrome, local tumors, schwannomas, metastasis, sarcoidosis, and Collet–Sicard syndrome.

A lesion at the cervicomedullary junction produces a weakness of the ipsilateral sternocleidomastoid and weakness of the *contralateral* trapezius.

*Outside of the skull:* Injury to the neck: Biting, blunt trauma, carotid endarterectomy, coronary bypass surgery, radiation, shoulder

blows, shoulder dislocation, stretch/hyperextension injury, strangulation, variants of neuralgic amyotrophy.

## Combination with Other CN

CN IX, X in base of the skull lesions or tumors.

## Causes and Frequency

*Dystonia:* A cervical lesion of the CN XI can result in cervical dystonia or torticollis (in addition to the more common cause of centrally caused dystonia).

*Iatrogenic:* Surgery in the neck (posterior cervical triangle), deep cervical lymph node removal, “neck dissection procedures,” shunt implantation (Fig. 16.2), fibrosis following radiotherapy, shoulder support in the Trendelenburg position.

*Neoplastic:* Collet–Sicard syndrome, ENT tumors, base of skull metastases (all tumors, in particular multiple myeloma, prostate, ENT, and



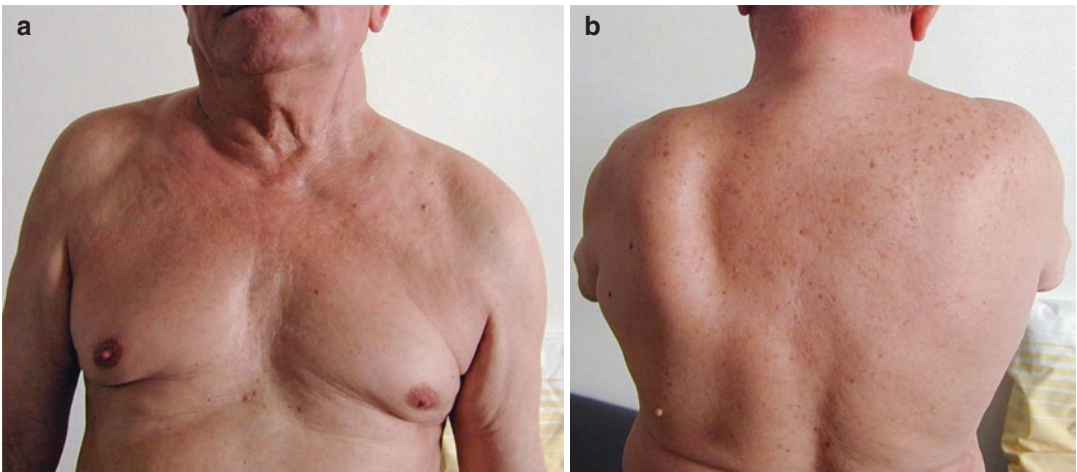
**Fig. 16.2** Accessory lesion below the left sternocleidomastoid muscle (after a shunt procedure). 1 atrophy of the trapezius muscle, 2 prominent difference in shoulder rounding, lower position of clavícula

Hodgkin's disease). Neurolemmoma, nerve sheath tumors. Spinal tumors, retrograde infiltration from adjacent tumors [2].

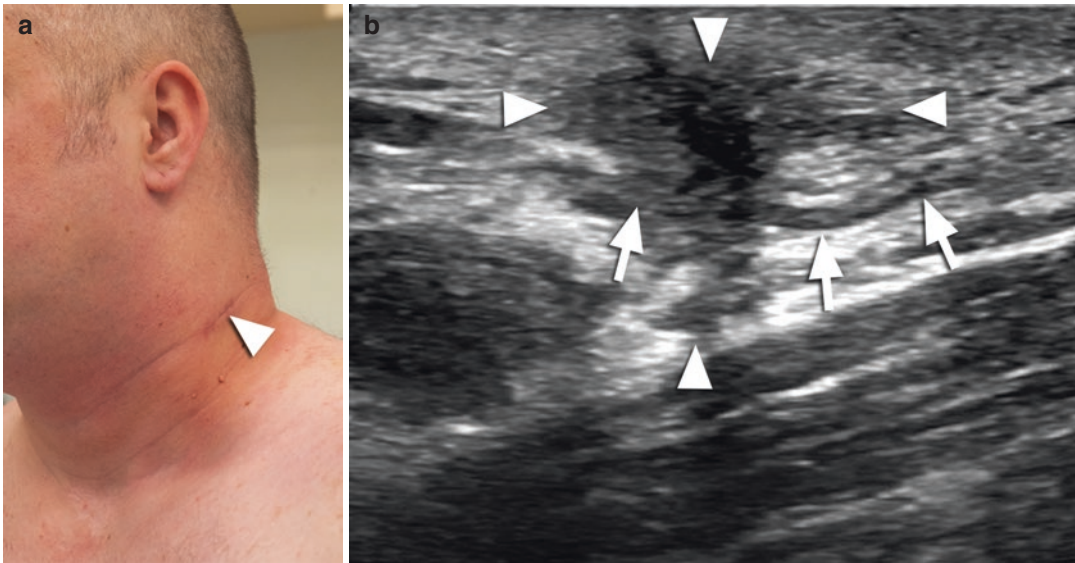
*Torticollis:* [3].

*Trauma:* [4], strangulation [5]. War and combat: Blunt and penetrating injuries to the neck, fractures of the jugular foramen.

*Others:* Motor neuron disorders, neck surgery (Fig. 16.3); spinal tumors and syringomyelia.



**Fig. 16.3** Left accessory nerve palsy, following carotid resection: (a) note the unilateral atrophy of the trapezoid muscle and (b) the winging of the scapula with the abduction of the medial scapular border



**Fig. 16.4** Ultrasound of the accessory nerve. (a) A complete loss of function of the trapezius muscle occurred after diagnostic surgical lymph node removal. (b)

Ultrasound examination revealed a scar tissue formation (arrowheads) surrounding the accessory nerve (arrows), which is intact

## Main investigations

Clinical and electrophysiology diagnosis.

Sternocleidomastoid muscle: Impaired head rotation.

Trapezius muscle: Upper, middle, and lower parts of the trapezius muscle must be examined separately. Upper and middle part lesions may produce winging of the scapula.

NCV: Stimulation of the nerve at the posterior aspect of the sternocleidomastoid muscle.

EMG: Sternocleidomastoid, trapezoid upper, middle, and lower part.

Imaging: MR of neck and shoulder muscles.

Ultrasound: The sternocleidomastoid muscle can be visualized in ultrasound (Fig. 16.4).

## Therapy

Nerve grafting (bridge): Reconstruction of the spinal accessory nerve with [6]; operations are not effective in long-standing scars; orthotic devices are not effective. The nerve is also used as a transferable nerve in neurotization and reinnervation [7].

Prognosis: Uncertain; recovery is slow and often incomplete. Further exploration is warranted if no improvement occurs after closed trauma.

## References

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