The Collet-Sicard syndrome as a complication of cardiovascular surgery

Multiple cranial nerve palsies are a diagnostic challenge as the nerves might be affected at any site along their course. A patient is described with a reversible lesion of the lower four cranial nerves due to cardiac surgery.

A 61 year old woman underwent cardiac surgery for a patent ducus arteriosus with mitral valve insufficiency. She had considerable left-right shunting with elevated pressure in the right side of the heart, resulting in tricuspid valve insufficiency. During surgery, a strongly blunted thoric aorta, aortic left subclavian artery and ductus arteriosus were discovered. The aortic arch was delivered and clips were placed between the carotid artery and the left subclavian artery as well as on the left subclavian artery. In the postoperative period, she suffered from aspiration pneumonia several times.

Six weeks after surgery, she developed speech and swallowing difficulties. Before surgery, the neurological examination had been unremarkable, but she now showed dysarthria, hoarseness and dysphagia. The function of the first eight cranial nerves was normal on both sides but the pharynx showed asymmetry on the left side with some numbness in that area. The pharyngeal constrictors were displaced to the right on phonation. Considerable tongue atrophy was present on the left side; on protrusion the tongue deviated to the left (fig). The left vocal cord showed paresis. The left sternocleidomastoid muscle showed paresis and atrophy. The trapezius muscle had normal function. There was no clinical evidence of autonomic dysfunction. Otherwise the neurological examination was normal.

The clinical diagnosis was the Collet-Sicard syndrome (CSS), a unilateral lesion of the last four cranial nerves, due to ischaemia in the territory of the neurosensineal trunk of the ascending pharyngeal artery (APA). Extensive blood and urinary examinations only showed a slightly elevated erythrocite sedimentation rate (ESR) and leucocytosis as a result of recurrent pneumonia. CSF examination was normal. CT scans, skull x-rays, also of the foramina, especially the jugular foramen, and chest x-rays were normal. Electromyography showed denervation and reinnervation activity in the left side of the tongue and the left sternocleidomastoid muscle. The signs and symptoms diminished and after one year, the neurological examination was normal.

A syndrome consisting of a unilateral lesion of the last four cranial nerves was described by Collet and Sicard and is now called the Collet-Sicard syndrome. For the differential diagnosis, a brainstem syndrome, the Villaret syndrome, and the cervical internal carotid artery dissection are relevant. A brainstem syndrome could definitely be excluded by physical examination. The Villaret syndrome consists of a unilateral lesion of the last four cranial nerves together with an ipsilateral incomplete Horner's syndrome with miosis and slight ptosis. This syndrome is generally caused by a mass in the retromandibular space, especially carcinomas and sarcomas behind the parotid gland extending into the parapharyngeal space. Our patient, however, did not have Horner's syndrome. Cervical internal carotid artery dissection might result in multiple cranial nerve dysfunction often accompanied by Horner's syndrome and mostly neck pain; our patient had no pain or autonomic dysfunction.

Lapresle et al reported on a patient who had a reversible vascular episode resulting in multiple cranial nerve dysfunction, probably due to catherisation of the APA during an attempt to reach the distal external carotid artery; nine months later, this patient showed almost complete spontaneous recovery. Devoize et al described four patients with paralysis of the lower four cranial nerves due to accidental or therapeutic embolisation in the APA during angiography. All the patients recovered or showed substantial regression of the deficits within six to nine months.

The vascularisation of the distal cranial nerves has been studied by Lasjaunias and Doyan and Lapresle and Lasjaunias. The APA arises from the external carotid artery and supplies the last four cranial nerves. The eleventh nerve receives dual vascularisation from the jugular as well as the musculospinal subdivision of the posterior branch of the APA, which explains why the nerve is sometimes spared in pathological events involving the APA. The trapezius muscle is not often involved in the CSS as was the case in our patient. Sufficient vascularisation is provided by the fact that the APA musculospinal subdivision also forms an anastomosis with the ascending cervical artery which supplies the middle cervical nerves.

The Collet-Sicard syndrome is a rare complication of radiotherapy to lumbar fields, and previous reports have described its onset from four months possibly up to 13 years following irradiation, though detailed clinical information was not provided. We report a case where symptoms began 23 years after irradiation for testicular neoplasia.

In December 1964 a 26 year old electrician had decision biopsy of a testicular teratoma which had been present for two months. Histology revealed testicular teratoma and he received cobalt irradiation to pelvis, para-aortic nodes and scrotum in thirty six fractions over seven weeks to a maximum dose of 4500 rads (estimated total dose to lower end of spinal cord and cauda equina—4920 rads). Chest x ray and abdominal examination revealed normal thereafter, but he developed a dusky skin reaction at the site of radiotherapy which was treated with topical emollients. He was followed up for nine years with no signs of recurrence. In 1973 he developed hypertension which was controlled on aldactone.

In April 1988 he developed a slowly progressive predominantly distal leg weakness which were marked on the right side. He revealed coarse fasciculations in both calves and right quadriceps. In the left leg there was mild weakness of knee flexion and extension and moderate weakness of all movements at the ankle. Power in the right leg was normal. Knee jerks were bilaterally brisk, but ankle jerks were depressed on the right and absent on the left. Sensation was normal and general