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 duc tus 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 thoracic aorta, aortic root, 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 unemongenal trunk of the ascending pharyngeal artery (APA). Extensive blood and urinary examinations only showed a slightly elevated erythrocyte sedimentation rate (ESR) and leucocytosis as a result of recurrent pneumonia. CSP 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 retroanterior plate, especially carcinomas, lymphomas 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. A fairly good recovery, as was the case in our patient, has been mentioned before.

Lapresle et al reported on a patient who had a reversible vascular episode resulting in multiple cranial nerve dysfunction, probably due to catheterisation of the APA during an attempt to reach the distal external carotid artery; nine months later, this patient showed almost complete spontaneous recovery. Dovéze 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 vasculatisation 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 vasculatisation 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 vasculatisation 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.

... Figure Six weeks after surgery the patient showed considerable tongue atrophy on the left side with deviation to the left on protrusion (A); after one year, the neurological examination was unremarkable (B).

Late onset radiation-induced motor neuron syndrome

Radiation-induced lumbosacral lower motor neuron 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 a diagnostic biopsy of a left inguinal testicle 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 remained 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 sign 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 was marked on the left. He also 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
A 63 year old man with a history of treated hypertension from 1979 had a myocardial infarction complicated by a left ventricular aneurysm and cardiac failure in 1981. In the early months of 1991 he had episodes of intermittent mild confusion and bifrontal headaches. These were felt to be related to low cardiac output, but were accompanied by stage heart failure and he received cardiac transplantation in May 1991 at which he was started on prednisolone, azathioprine (10 mg daily) and cyclosporine (initially given intravenously but changed postoperatively to 3 mg/Kg/day orally, a low dose being used because of his slightly impaired renal function), nystatin, ranitidine, thiamine and aspirin. There was a marked exacerbation of confusion without localising features which progressively increased. A CT brain scan one month postoperatively showed deep white matter changes and a small left parietal infarct. Two days after this he began to have episodes of dysphasia and right sided weakness lasting from 30 minutes to several hours with full recovery between episodes. He was transferred to Charing Cross Hospital for further assessment. On arrival he was unresponsive and exhibited version of head and eyes to the right with shaking of the right arm and facial grimacing. He was treated with intravenous diazepam and attempted to drum on his right leg. His right focal fitting, allowing him to recover consciousness and speak when he was immediately orientated, able to give his name, the date and date of birth and obey three step commands. There were no focal neurological signs. The cyclosporine dose was reduced to 2 mg/Kg, he was given a phenytoin load and the diazepam maintained as an infusion for 18 hours. Carbamazepine was introduced 12 hours after admission because of persisting focal fitting.

Full blood count revealed a haemoglobin of 11.0 g/dl with normal indices, a white cell count of 11.8 x 10^9/l and a platelet count of 273 x 10^9/l. Biochemical evaluation showed normal sodium, potassium and glucose, a urea of 9.9 mmol/l, creatinine 145 mmol/l, bilirubin 1.3 mmol/l and alkaline phosphatase 276 U/l but urate, proteins, aspartate aminotransferase, calcium and phosphate were all normal. Magnesium in serum was 0.74 mmol/l and in red cells 1.9 mmol/l (normal range in red cells 1.8 to 2.7 mmol/l). The CSF contained no cells, a protein of 0.5 g/l and glucose of 4.8 mmol/l. Repeat CT scan revealed no new abnormalities. Serum cyclosporine levels were 50 ng/ml on admission and 105 ng/ml three days later (therapeutic range 75–150 ng/ml in serum) despite the dose reduction.

As the focal fitting abated at 24 hours post admission frequent bilateral myoclonic jerks became apparent which persisted for a day after the fitting ceased. At this stage his higher intracranial functions were rapidly retrograded to normal. He was readmitted to hospital on the third day and after a second CT scan had been performed was referred to the neurological ward. He was said to feel uninterrupted but could not follow commands and had caused a sore throat and explained the difficulty he had had with speech. He felt that his throat was still involved for several minutes. He was referred to the neurological ward and after a second CT scan had been performed was referred to the neurological ward.
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