The role of chronic superior caval syndrome and stenosis of jugular veins in neurodegenerative diseases. Surgical treatment and preliminary results

Chronic superior caval syndrome (CSCS) and stenosis of jugular have been suggested to play a role in the pathogenesis of several degenerative disorders of the central nervous system. Although controversy still remains as to whether anatomic and/or functional alterations of the cerebrospinal venous effluent really contribute to the development of the disease. Several reports have shown that restoration of a normal venous flow pattern by internal jugular veins (IJV) angioplasty (PTA) can improve neurological status and functional capacity. It is thought that in the event of a stenosis of the superior vena cava, the cerebrospinal venous circle normally flows into the jugular veins and brachiocephalic veins and, by means of the superior intercostal veins and the mammary veins, it reaches the azygos and inferior vena cava. Recent studies have demonstrated that in the presence of a stenosis of the vena cava or of the brachiocephalic or the jugular veins, venous blood can invert the direction of its flow and move towards the cerebrospinal circle. The cause of the reflux has been attributed to increased central venous pressure and volume overload. Assuming that in the venous system a countercurrent is not possible, the direction of the flows is always monodirectional and centripetal. The fundamental difference between the cerebrospinal venous circulation (CVS) and the systemic venous system is the lack of valid bidirectional venous flow.

The concept of bidirectional flow is extremely important. The Cerebrospinal Venous System (CVS) in the absence of valves, allows to provide a pressure homeostasis to the cerebral circulation. The CVS, as already described by Batson, Epstein and colleagues, has a large capacitance, such as a lake-venous. Blood flows into the brain and from the brain, depending on the patient's posture or intrathoracic and intra-abdominal pressure. The Cerebrospinal Venous System (CSVS) extends from the head to the pelvis, a group of veins and venous plexuses without valves in close communication.

The CSVS is divided into two parts:
1. Intracranial veins including cortical veins, dural sinuses, cavernous sinuses and ophthalmic veins
2. Vertebral veins and its venous plexuses along the vertebral column

Vertebral veins and intracranial veins join in the occipital region. At the lumbar level the Cerebrospinal Venous System communicates with the sacral, pelvic and prostatic plexuses. The main pathway of cerebral blood outflow is the extracranial venous system through the internal jugular vein anteriorly and the azygos system posteriorly. From the sigmoid sinuses, the blood reaches the internal jugular vein and then the superior vena cava through the brachiocephalic vein. The vertebral venous system is connected with the deep veins of the thorax and vertebral column, the intercostal veins, the azygos vein and the emiazygos to reach the superior and inferior vena cava. The anatomo-functional characteristics that we have described, have allowed us to hypothesize and subsequently demonstrate with angiography that, in the superior caval syndrome, the Cerebrospinal Venous System is a compensation circle, which can directly connect the upper cavity with the inferior vena cava. The hypothesized mechanism in the superior caval syndrome is determined by the venous flow in obstruction, creating a pressure gradient between the right atrium and the superior vena cava. The flow is reversed, from the internal jugular vein to the cranial sinuses, the cavernous sinuses, the venous plexus of the azygos and the emiazygos, then the superior vena cava, under the obstruction or the inferior vena cava and finally in the right atrium. In addition to an increase in blood pressure, volume overload is also established due to the large amount of blood in the upper limbs, chest and head.
The clinical situation that emerges is very unpredictable. The symptoms are many and the pathophysiological mechanisms responsible are an increase in cerebrospinal pressure and a volume overload. These two mechanisms cause:

1. A slowing of the microcirculation with impaired cellular metabolism for the reduction of the supply of oxygen and nutrients and difficult removal of carbon dioxide and catabolites.

2. Reduced venous drainage in the brain resulting in stasis of the venous circulation and increase in the volume of the cerebral ventricles.

3. A possible passage of toxic substances such as bacteria, viruses, tumor cells from the peripheral tissues to the brain.

The main neurological manifestations are:

1. Fatigue, the most common symptom (85% of patients)
2. Hyposthenia to one or more limbs, paresthesias, headache, sleep disorders, cognitive disorders, diplopia or reduction of the visus, balance disorders, micturition disorders, dysphagia.

The physiopathological knowledge and skills acquired in the surgical treatment of the superior vena cava syndrome, with internal angioplasty of the jugular vein, have allowed us to extend this surgical procedure, even in those patients with chronic neurodegenerative disease, such as sclerosis multiple, Parkinson’s disease, Meniere’s syndrome, which has had a temporary but documented benefit after an angioplasty. Vessel patency was re-established by removing before all valve anomalies and thrombotic material and by applying an autologous saphenous vein enlargement patch.

Preliminary results:

There are 128 patients undergoing surgery from May 2011 to March 208. Follow-up is from 2 to 84 months. The long-term vessel patency with saphenous vein patch is 96% documented with an venous doppler eco scans. After surgery, all patients underwent an intense rehabilitation program that resulted in a progressive improvement of the clinical and neurological picture. To prevent thrombotic complications, they were maintained on oral anticoagulants (warfarin) for six months, with a target INR of 2-3. They were then started on aspirin 100 mg daily. No early or late complications were observed. Throughout the observation period patients were maintained on immunosuppressant agents. Follow-up visits were performed three, six and twelve months after surgery. Venous Doppler Eco scans confirmed optimal vessel patency with a normal venous flow pattern. Concurrently, the neurological status assessed by the Expanded Disability Status Scale (EDSS) and Barthel Activities of Daily Living Index progressively improved along with reduction of fatigue (MFIS Scale), increased muscular strength and improved balance (Newberg Scale). In our experience, we considered the caliber of jugular veins and the number of brain lesions on MRI and we divided the patients into three groups: In the first group, patients with a good caliber of the internal jugular vein, reduced number of brain lesions on magnetic resonance and with good surgical result. In these patients, there has been a noticeable and constant clinical improvement over time with recovery of a good quality of life, no remote recurrence (28 cases). In the second group, patients with a good caliber of the internal jugular vein, multiple brain lesions on magnetic resonance but with good surgical result. These patients showed an improvement in symptomatology, but poor variation in symptoms related to brain injuries (92 cases). The third group, patients with small-vein veins and with poor development of brain veins. These showed no improvement in symptomatology (9 cases). Currently these patients are no longer selected for the surgical procedure. In consideration of the results obtained, the proposed surgical procedure, enlargement of jugular veins with saphenous vein patches may be considered the treatment of choice in the superior caval syndrome associated with jugular vein stenosis in association with chronic neurodegenerative disease. Indeed, according to our previous and present experience, surgery is almost invariably feasible and grants durable results in terms of vessels patency. The concurrent improvement in the clinical and neurological picture consistently observed in our patients further supports the idea that, at least in some patients, restoration of normal venous flow by surgical repair may contribute to limit disability and disease progression. Successful surgical repair re-established long-term vessel patency and significantly improved neurological symptoms.

Biography

Salvatore Spagnolo currently working in Cardiosurgery (ICLAS - Istituto Clinico Ligure di Alta Specialty) Gvm Care & Research, Italy. His wide range of publications in various national and international journals

spagnolo.salvatore@libero.it