Videoendoscopic Nasal Polypectomy In Churg-Strauss Syndrome: Anaesthesiological Management

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Citation

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Abstract

Churg-Strauss syndrome (CSS)isan allergic or autoimmune reaction to an environmental agent or drug, consisting of the allergic, the hypereosinophilic, and the vasculitic stage. Related to anesthesia, asthma and pulmonary infiltrates are adverse conditions for a general anesthesia, whereas neuropathies and neuritis can be contraindications to neuroaxial and peripheral anesthesia.
Case: 61-year-old woman undergoing an intervention of nasal polyposis videoendoscopical resection.
Anamnesis: Churg-Strauss vasculitis with multineuropathy, nephropaty, ischemic hearth disease, hypertension, obesity, hypothyroidism, osteoporosis, allergic asthma, nasal polyposis, stenosis and hypo-osmia.
Anaesthesia: general balanced narcosis, with antiallergic prophylaxis. Throughout the surgical intervention: steady-state of cardio-respiratory parameters, pulmonary pressure and compliance. Awakening, extubation, and the postoperatory presented any complications; pain control was good.
Literature doesn't offer many guidelines for proper anesthesiological conduct in CSS patients: preoperative examinations, optimizing of antiallergic therapy and antiasthmatic prophylaxis are fundamental in the management of general anaesthesia in such patients.

INTRODUCTION

In 1951, Churg and Strauss first described the syndrome in 13 patients with asthma, eosinophilia, granulomatous inflammation, necrotizing systemic vasculitis, and necrotizing glomerulonephritis: this rare systemic autoimmune disease is characterised by inflammation of small- to medium-sized arteries, arterioles and venules: this inflammatory process of blood vessels is more commonly known as vasculitis. It can affect people of all ages with the average age at diagnosis being 35 to 45 (men slightly more than women). Estimates about the incidence of CSS vary widely and range from 2.4 to 10 cases per 1 million people, or roughly from 720 to 3,000 people in the United States.

Causes of Churg-Strauss syndrome are unknown. Churg-Strauss syndrome is possibly an allergic or autoimmune reaction to an environmental agent or drug. Several case reports have described drug-induced forms of Churg-Strauss syndrome. Mesalazine-induced Churg-Strauss syndrome has been reported in a patient with Crohn disease and sclerosing cholangitis (Sinico, 2006); 4 publications have addressed the association between propylthiouracil, methimazole, and vasculitides, including Churg-Strauss syndrome. One report is available on the association of freebase cocaine and Churg-Strauss syndrome (Orriols, 1996).

CSS is a progressive disease consisting of three distinct phases: the allergic, the hypereosinophilic and the vasculitic stage (even though an early diagnosis and an effective treatment can prevent the disease from progressing to the last stage).

The allergic phase is almost always characterised by asthma. People who have had asthma throughout their lives experience a worsening of symptoms that become more difficult to treat. Sinus disease, which is characterised by facial pain from sinusitis, nasal polyps, allergic rhinitis (inflammation of the mucous membranes of the nose causing sneezing, itching, runny nose), and recurrent pneumonia and/or bronchitis are also typical of this prodromal phase. This phase can last from 4 to 27 months, although some patients stay in this phase for many years.

The second phase is called hypereosinophilic. This overabundance of eosinophils can occur either in the blood or in the tissues: during this phase, patients often suffer from chronic eosinophilic pneumonia and eosinophilic

gastroenteritis (inflammation of the esophagus, stomach or intestines). Symptoms can include weight loss, fever, and night sweats. In addition, if lungs are affected, patients may experience shortness of breath, a feeling of heaviness in the chest, and a constant cough. Should the gastrointestinal tract be involved, patients may experience abdominal pain, bloating, vomiting, diarrhea, and nausea, sometimes resulting in weight loss and even anorexia.

During the systemic vasculitis phase, there may be inflammation and damage to blood vessels throughout the body; this, in turn, may damage many different organs, and symptoms vary widely depending on the organ affected. Organs mainly affected by CSS include skin (rashes, purpura, urticaria), heart (pericarditis, chest pain), lungs (hemoptysis), central nervous system (intellectual or motor disturbances), peripheral nervous system (peripheral neuropathy, motoneuritis), gastrointestinal tract (diarrhea, vomiting, nausea) and less commonly, kidneys, eyes and musculoskeletal system.

When CSS is diagnosed early on, patients may suffer from asthma and tissue eosinophilia without detectable vasculitis: they may have eosinophilic pneumonia and/or lymphnode involvement.

Figure 1

Figure 1: Nasal Polips

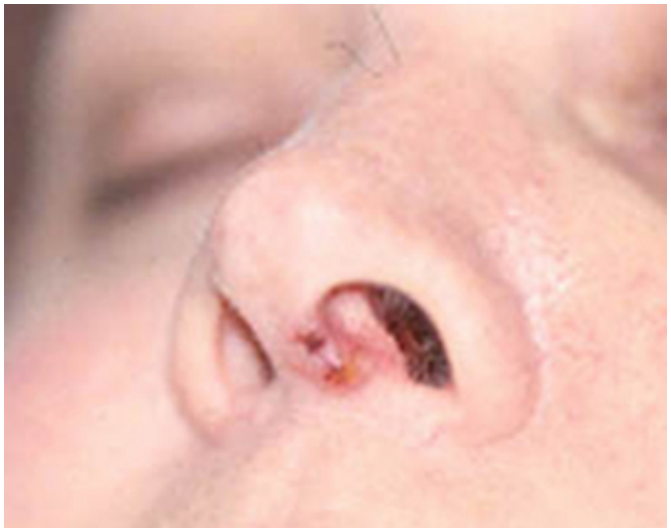


Figure 2

Figure 2: Nodules

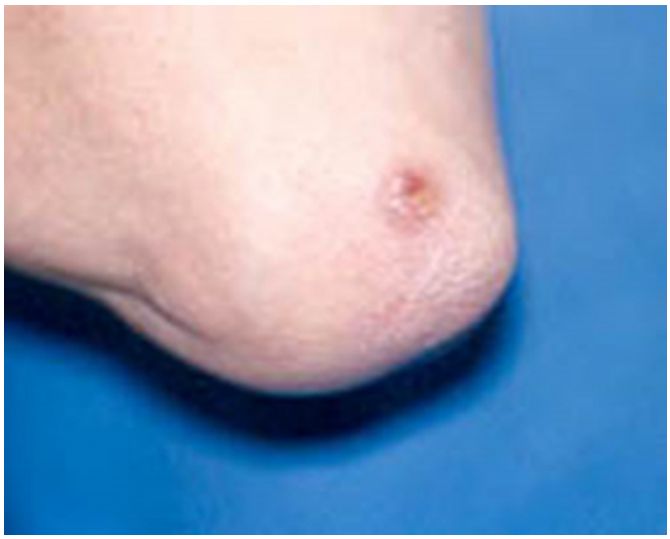


Figure 3

Figure 3: Skin Rashes

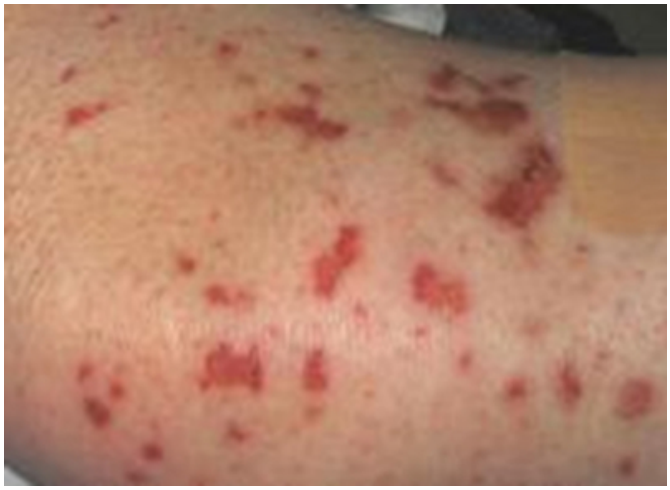
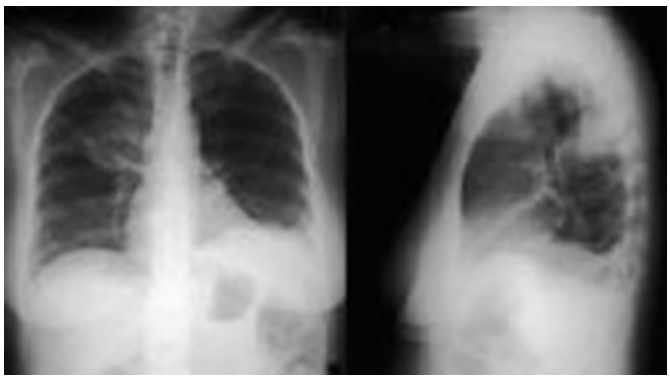


Figure 4

Figure 4: Pulmonary Infiltrates



In 1990, The American College of Rheumatology (ACR)

established six criteria that distinguished Churg-Strauss Syndrome from other vasculitic diseases. These are: history of asthma; eosinophilia > 10% on a white blood cell differential count; mononeuropaty or polyneuropaty; fixed or transitory pulmunary infiltrates; history of paranasal sinus abnormality; presence of extravascular eosinophilia. The presence of four or more of these six criteria yielded a sensitivity of 85% and a specificity of 99.7%. The disease at this early stage can often be treated with steroids (Prednisone, Medrol), instead of more damaging chemotherapy (Cyclophosphamide): this is why an early diagnosis is so important. Unfortunately, most of the treatments for CSS have severe side effects. The key is to try and maintain on as low a dose of medication as possible to avoid side effects.

Prior to the advent of prednisone, CSS was often a fatal disease. The majority of patients died from rampant, uncontrolled disease. With present therapy, constitutional symptoms begin to resolve quite quickly, with gradual improvement in cardiac and renal function, as well as improvement in the pain that results from peripheral nerve involvement. The course of therapy can last for 1 to 2 years, although the length and type of treatment depend on the severity of disease and the organs involved. The patient's response to treatment and the continuation of disease control during lowering of the prednisone dose are the primary determinants of how long therapy is continued. Laboratory monitoring of blood tests is very helpful in gauging the activity of disease. Some of the most useful laboratory tests are the erythrocyte sedimentation rate (ESR) and the eosinophil count.

In literature only two cases of patients who suffer from Churg-Strauss syndrome, showing a cholinesterase deficiency despite normal phenotypes, have been reported: during the immunosuppressive therapy they have both suffered from prolonged paralysis in ICU.

Regarding this pathology related to either general or local anesthesia, there need to be considered different symptoms as contraindications: asthma, pulmonary infiltrates and a positive anamnesis for allergies are adverse conditions for practicing a general anesthesia; whereas mononeuropathies, and mono- and polyneuritis can be contraindications to practicing neuroaxial and peripheral anesthesia. At any rate, patients showing such pathology need a thorough preoperatory analysis taking advantage of examinations and a further specialist consultation aiming to focus the stage of the disease and the condition of the patient: in the event of a general compromised situation, we have to consider the necessity of a postoperatory admission under Intensive Care Unit.

The case reported observes perioperative management of a patient affected by such pathology and subject to a surgery of videoendoscopy ablation of nasal polyposis, carried out under general anesthesia.

CASE REPORT

The case analysed concerns a 61-year-old woman familiar with ischemic heart disease and undergoing an intervention of nasal polyposis videoendoscopical resection. Previous surgery: appendicectomy at the age of 20 and hysterectomy with bilateral annessiectomy at the age of 48, non smoker, physiological bowel habit and diuresis. Her anamnesis presented Churg-Strauss vasculitis diagnosis with multineuropathy (supported by biopsy of sural nerve) and nephropaty, hypertension, obesity, hypothyroidism, osteoporosis, allergic asthma, nasal polyposis with stenosis and hypo-osmia: such a complicated clinical condition required various highly specialised preoperatory examinations.

The neurological examination assessed 5/5 strength in right arm, 4 ½ in left arm, 5- in right leg and 4 ½ in left leg; weak/lacking DTR in all four limbs, Babinski's sign negative on the left, and neutral on the right; paresthesias of finger I and II of the right arm, finger IV and V of left arm, distal hypoesthesia to left elbow, right knee and right foot. Slight postural and rest tremor of distal upper limbs; isochoric, isocyclic and normally reactive pupils, apparently non injured cranial nerves, non meningeal symptoms, able to walk without a support but with a clumsy gait.

The rheumatologists examination assessed a good general level of inflammation index and a constant sensory neuropathy in the sural nerve, previous biopsy site (on regenerative basis?); nephrologic assessment analysed organ function (creatinine level 1,1 mg/dL, BUN 23, clearance level 65 mL/min, absent proteinuria and slight microematuria were observed).

The examination for allergies tested pollen, perennial inhalant, Mycophyta and latex allergens; the patient had positive skin prick test results to Dermatophagoides and the consultant recommended RFT, considering that the patient's positive anamnesis had revealed allergic asthma: basal spirometric values (FVC 2,86; FEV_1 2,42; FEV_1/FVC 78%) and DLCO values were within levels accepted; the patient

suffered also from acetylsalicylic acid and furosemide intolerance.

Rhinosurgical case history showed nasal obstruction, congestion, posterior rhinorrhea, and hypo-anosmia as major symptoms of polyposis; frontal cephalea was a minor symptom with temporal ratio chronic-persistent > 12 weeks. Anterior septal deflection, inferior turbinates hypertrophy with purulent exudates, and middle meatus polyposis with chronic rhinosinusitis were detected.

Current therapy:

- Mofetil mycophenolate 1000 mg/die
- Metolazone 5 mg/die
- Amlodipine 10 mg/die
- Spyronolactone 25 mg/die
- Lansoprazol 20 mg/die
- Levothyroxine 50 mcg/die
- Salmeterol aerosol 50/100 mg/die
- Pregabalyn 450 mg/die
- Calcium/Vit. D 1 pill/die

Anthropometric parameters and preoperatory clinical examination: height 165 cm, weight 93 kg, sensorium conscious and oriented; thyro-hyoid distance, mental-thyroidal distance in head extension, as well as interincisor distance shortened; Mallampati test II/III, removable dental prothesis along the inferior arcade (such parameters implied a predicted difficult intubation). ECG results: sinus rhythm, left axis deviation, no significant alterations in repolarization; blood pressure 145/85 mmHg, pulse 77 bpm, negative cardiologic clinical examination, no peripheral stagnation oedemas; chest X-ray disclosed an anterior disventilatory band in middle area on the right lung; during auscultation, low intensity breath sounds over the whole area were detected.

Altered blood test, only: RDW: 15%; fibrinogen: 4,55 g/L; magnesium: 1,5 mg/dL; CPK: 250 U/L; total cholesterol: 226 mg/dL; albumine: 46 g/L.

The evening before surgical intervention, intravenous antiallergic prophylaxis was administered to the patient. The treatment was Prometazine Cloridrate 50 mg, Betamethasone disodium phosphate 4 mg, Ranitidine chlorhydrate 50mg dissolved in 100 mL of a physiological salt solution. The same treatment, combined with antiobiotic prophylaxis, was administered 30 minutes before the injection of anesthesia. Peripheral venous access being found with a needle catheter 18 Gauge, Fentanyl 0,1 mg, Propofol 200 mg, Succinylcholine chloride 90 mg were injected. Orotracheal intubation was practiced with a rigid fiberscope under indirect vision. Afterwards, 18 mg Cisatracurium besilate were administered. Intra-operative anesthesia was maintained with MAC 0,7 – 0,8 inhalation of Sevoflurane, combined with a mix of O_2 (40%)/N_2O (60%) and continuous intravenous infusion of Remifentanil (variable dosage ranging from 0,5 to 2,4 mcg/kg/min): surgical duration was 3 hours 20 minutes. About 20 minutes before the patient's awakening, 100 mg of Tramadol, 50 mg of Ranitidine and 4 mg of Ondansetron were dissolved in 100 mL of a physiological salt solution and then administered by intravenous injection. Throughout the whole surgical intervention, a steady-state of cardio-respiratory parameters and no significant alterations in pulmonary pressure and compliance were observed; the amount of blood loss during surgery was about 400 mL, and the patient was kept hydrated by 1000 mL of a 0,9% physiological salt solution and 500 mL of Lactated Ringer's solution. During the postoperative period, 1500 mL/24h of crystalloids, antithromboembolic prophylaxis with low-molecular-weight heparins for 10 days, and 1 g of Paracetamol (max 3/die) as a pain treatment – in the event of V.N.S. ≥ 5/10 – were prescribed.

Polypectomy with microdebrider, anteroposterior bilateral ethmoidectomy, and bilateral middle meatus antrostomy with drainage of mucopurulent secretion, were performed with nasal videoendoscopy. Afterwards, a bilateral sphenoidotomy with drainage of mucin, bilateral opening of ethmoidofrontal cavity, partial bilateral middle turbinectomy, application of hemostatic foam in ethmoidal cavity, and nasal tampon with Merocel were performed.

The awakening, the extubation, and the early postoperatory haven't presented any complications; pain control was good. The patient was brought to the ward 30 minutes after the end of surgical intervention. Not even additional complication occurred during postoperatory admission, and the patient was discharged within the scheduled time.

During postoperative admission, the patient was subject to basal hematochemical examinations; thyroid hormone dosage, and examinations to assess the renal function, as

well as to neurological and pneumologic check-ups; the condition of the patient was alike the preoperatory one, and so remained one month after surgical intervention.

DISCUSSION

In the literature, it is rare to find patients suffering from Churg-Strauss syndrome undergoing anesthesia: Guryar M. et al. explain the perioperatory management of a patient subject to radical mastectomy under combined anesthesia (general and epidural); whereas Ito Y. et al. and Sato H. et al. relate two cases of patients suffering from severe asthma, respectively subject to subdural and to general anesthesia, in which steroid therapy has well controlled such pathology. Two episodes of cholinesterase deficiency are reported by Taylor BL. et al., who illustrate one aspect of the pathology – observed only empirically, and whose physiopathological basis are still to be studied and proved further.

As observed, literature doesn't offer many guidelines on which a proper anesthesiological conduct can be set up regarding patient suffering from such pathology. Thus, it is fundamental a preoperative study of the pathology, focusing also on grading, pathological prevalence, and therapy optimization: in this area, it is vital to take advantage of consultation and multispecialist examinations. In patients undergoing general anesthesia, it is crucial, above all, to perform an antiasthmatic therapy optimization, in order to avoid perioperatory perils. In the case reported none of such difficulties has occurred, so it is implied that preoperatory antiasthmatic prophylaxis and anaestesilogical premedication has been effective.

Moreover, it is just as important a thorough analysis of kidney function, in order to correctly measure a patient's ability to metabolise many anesthetic drugs, and to determine his/her cardiac condition, considering the presence of hypothetical pericarditis. Whereas, regarding patients subject to regional blockade (central or peripheral), there need to be carried out a careful analysis of neurodegenerative aspects of the pathology, in order to administer anesthesia without taking any risk. Anyway, regarding such patients, there has to be an admission under Intensive Care Unit available during the early postoperatory, should severe – especially respiratory – complications occur.

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