Obstetrical and Pediatric Anesthesia

Anesthetic management of the parturient with relapsing polychondritis

[Anesthésie d'une parturiente atteinte de polychondrite chronique atrophiante]

M. Joanne Douglas MD FRCPC, Stephanie Ensworth MD FRCPC

Purpose: To present the anesthetic management of a parturient with relapsing polychondritis (RP) and to discuss the anesthetic implications of RP.

Clinical features: A 28-yr-old primiparous woman with known RP, spondyloarthropathy and fibromyalgia presented for urgent Cesarean delivery for breech presentation and prodromal labour. Her pregnancy had been complicated by a hospital admission for an exacerbation of her RP as manifested by hoarseness, increased pain and tenderness of her left ear and nasal bridge cartilages, sinusitis with bloody nasal discharge and increased pain and tenderness of the anterior tracheal rings. Epidural anesthesia was administered for the Cesarean delivery. Her intraoperative and postoperative course was uneventful. Close cooperation among obstetricians, anesthesiologists and rheumatologists resulted in a successful outcome.

Conclusion: Relapsing polychondritis is a syndrome with important anesthetic implications. Multidisciplinary cooperation is essential in managing these high risk parturients.

Objectif: Présenter l'anesthésie d'une parturiente atteinte de polychondrite chronique (PC) et discuter des implications anesthésiques de la PC.

Éléments cliniques: Une primipare de 28 ans, suivie pour PC, spondylarthropathie et fibromyalgie, a été admise pour une césarienne d'urgence à cause d'une présentation du siège et d'un faux travail. La grossesse s'était compliquée d'une hospitalisation pour exacerbation de la PC manifestée par la raucité de la voix, une douleur et une sensibilité plus grande de l'oreille gauche et des cartilages de la racine du nez, une sinusite avec écoulement nasal sanguinolent et une douleur et une sensibilité accrues des cartilages antérieurs de la trachée. La césarienne, réalisée sous anesthésie péridurale, a connu une évolution peropératoire et postopératoire

sans incident grâce à la collaboration étroite des obstétriciens, anesthésiologistes et rhumatologues.

Conclusion : Les parturientes atteintes de PC sont à haut risque et doivent être prises en charge par une équipe multidisciplinaire.

RELAPSING polychondritis (RP) is a rare condition of recurrent inflammation of cartilage and connective tissue.¹ Although it tends to occur in middle age (> 40 yr) it has been reported in younger individuals, including parturients. The literature provides some information regarding anesthetic management for the surgical treatment of complications of RP, but there are no reports describing the anesthetic management of the pregnant woman with RP. Ethical review for publication of this report was received from the University of British Columbia (UBC) and BC Women's Hospital. The subject gave written, informed consent for its publication.

Case report

The patient was a 28-yr-old primiparous woman who was previously diagnosed with RP,^{1,2} spondyloarthropathy and fibromyalgia.³ She had keratoconus, unrelated to her RP or spondyloarthropathy. She was referred to the anesthesia antenatal clinic at 20 weeks gestation to plan for her labour and delivery.

Her complex medical history dated to age 15 yr when she had intermittent episodes of hot, red, swollen and tender helices of her ears, as well as morning

From the Departments of Anesthesia and Medicine, Division of Rheumatology, B.C. Women's Hospital, University of British Columbia, Vancouver, British Columbia, Canada.

Address correspondence to: Dr. Joanne Douglas, Department of Anesthesia, B.C. Women's Hospital, 4500 Oak Street, Vancouver, B.C. V6H 3N1, Canada. Phone: 604-875-2158; Fax: 604-875-2733; E-mail: jdouglas@cw.bc.ca

Accepted for publication February 22, 2005.

Revision accepted May 10, 2005.

lumbar spine stiffness that lasted 30 min. At age 23, her spine stiffness increased and she was diagnosed with spondyloarthropathy but there was no radiologic evidence of bony spinal fusion. She was active physically and worked full time as a fitness instructor until age 24, when she went on to long-term disability. At age 24, she developed intermittent loss of her voice, tenderness over the bridge of her nose and fever leading to the diagnosis of RP. She also was diagnosed with fibromyalgia when she presented with widespread, non-specific muscular pains. Her respiratory function was followed by a respirologist; pulmonary function studies, bronchoscopy and computed tomography scan of her chest were all normal pre-pregnancy. Ten years prior to the current pregnancy she had received uneventful general anesthesia for surgery on her temporomandibular joint.

Medications in the past included prednisone and methotrexate. At the time of the anesthetic consultation at 20 weeks gestation her daily oral medications were: ibuprofen 1200 to 1800 mg, acetaminophen with codeine *prn*, folic acid, prenatal vitamins, aspirin 81 mg and calcium. At that time, the pregnancy had been relatively uncomplicated with no evidence of gestational hypertension, diabetes or coagulopathy. Erythrocyte sedimentation rate (ESR) was elevated at 100 mm·hr⁻¹ and the patient reported occasional palpitations although a previous 48 hr Holter monitor evaluation was reportedly within normal limits. Additional complaints included spondylarthritic chest wall pain, limited range of motion of her neck and pain and stiffness of her thoracic and lumbar spines. As well, she had tenderness over her thyroid and cricoid cartilages. She continuously had effusion, pain and stiffness of the small joints of her hands. At the time of the clinic visit, the rheumatologist considered her RP to be relatively mild and stable.

Near the end of the second trimester (estimated gestational age 24 weeks) her RP and spondyloarthropathy remained active as manifested by tenderness of the left ear helix, cartilaginous bridge of the nose and tracheal rings; swelling of her nasal mucosa; bilateral Achilles tendonitis with swelling; thoracolumbar spondylitis; sacroiliitis, anemia of chronic inflammatory disease (108 g·L⁻¹) and markedly elevated ESR to 100 mm·hr⁻¹ (Westergren). Due to her gestational age the ibuprofen was discontinued and prednisone 10 mg *po* daily and azathioprine 75 mg·day⁻¹ *po* were commenced.

At 33 weeks gestation the patient was admitted to hospital for an exacerbation of RP manifested by increased pain and tenderness in the left ear cartilage and nasal bridge, sinusitis with bloody nasal discharge,

increased tenderness and pain of the anterior tracheal rings and hoarseness of her voice. The spondylitis was also more active with six areas of enthesitis involving the right knee and both feet; pubic symphysis; anterior costochondritis and ongoing thoracolumbar spondylitis with sacroiliitis. The prednisone dose was unchanged at 10 mg·day⁻¹ *po*, but azathioprine was increased to 125 mg·day⁻¹ *po*. Her symptoms improved and she was discharged home after three days.

On that admission, physical examination showed a modified Mallampati class 1 airway, ability to protrude her mandible in front of her maxilla, and a normal thyromental distance. She had limited range of motion of her cervical spine and tenderness over the larynx and tracheal rings. There was tenderness to palpation of the small joints of her hands. She had limited ability to round her lumbar spine and her spinous processes were tender. The remainder of the physical examination was normal.

At 39 weeks gestation she was admitted with a five-day history of prodromal labour. As the fetus was breech and the cervix long and posterior, the obstetrician decided to perform a Cesarean delivery. Ranitidine 50 mg and metoclopramide 10 mg were administered *iv* and sodium citrate was given orally for antacid prophylaxis. In the sitting position, an epidural catheter was inserted easily at the L3–4 interspace, using loss of resistance to saline. Carbonated lidocaine (2%) with epinephrine 1:200,000 was administered incrementally to a total of 18 mL, along with 75 µg of epidural fentanyl. This provided a T4 level to pinprick bilaterally. She required an additional 9 mL of local anesthetic mixture intraoperatively due to prolonged surgery. Epidural morphine 3 mg was given for postoperative analgesia. Oxygen saturation, non-invasive blood pressure and heart rate (3-lead electrocardiogram) remained normal throughout surgery. Prophylactic antibiotics were administered as per hospital routine, and she received 2 L of normal saline prior to surgical incision. The estimated blood loss was 800 mL. A 3415 g male was delivered with Apgar scores of 6 and 9 at one and five minutes respectively. In order to prevent relative adrenal insufficiency she received hydrocortisone 150 mg *iv* 30 min preoperatively, 125 mg *iv* six hours postoperatively and methylprednisolone 50 mg *iv* the following morning. Azathioprine and prednisone were reinstituted when she was drinking fluids adequately.

The patient and her newborn were discharged on the fourth postoperative day. Her medications were oral azathioprine 100 mg·day⁻¹, ibuprofen 600 mg tid, omeprazole 20 mg·day⁻¹ and prednisone 10 mg·day⁻¹ which was to be tapered to 5 mg·day⁻¹ by

five weeks postpartum. Six weeks postpartum the tenderness of her nasal bridge and trachea had reportedly decreased.

Discussion

Relapsing polychondritis is a rare inflammatory disorder of unknown etiology, although some postulate an autoimmune mechanism.¹ RP is characterized by episodic inflammation with subsequent degeneration of cartilage and connective tissue, classically involving the upper airway.¹ Typically, Relapsing polychondritis presents with erythema, pain and swelling of cartilaginous structures such as the ears, nose, tracheal rings and various joints. The diagnosis is confirmed when the patient has three of the six features of the disease: bilateral auricular chondritis, nonerosive seronegative inflammatory arthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis and audiovestibular damage and histologic confirmation.¹ The subject of this case report fulfilled four of the six RP diagnostic criteria.^{1,2}

Laryngotracheal symptoms occur in approximately 25% of patients at the time of initial diagnosis, but ultimately these symptoms will occur in 50% of RP patients.⁴ Airway involvement is more common in women than in men (2.6:1) and indicates a poor prognosis.⁴ Respiratory failure due to collapse of various portions of the tracheobronchial tree is the most frequent cause of death in patients with RP. Inflammation can also involve other proteoglycan-rich structures such as the eye, heart, blood vessels and inner ear.^{1,2} The second most common cause of death is from cardiovascular involvement. Aortic or mitral valvular insufficiency occurs in a small number of patients.⁵ Arteries and veins of all sizes may be affected with vasculitis and when the aorta is involved, life-threatening aneurysms may result. Patients with tracheobronchial collapse, valvular dilatation or vasculitis may require surgical treatment (stenting; valve replacement or grafting of the affected vessel, usually the aorta).

Co-morbid diseases have been reported in conjunction with RP. These include the systemic vasculitides; spondyloarthropathy; connective tissue diseases; hematologic diseases; and other diseases. Our patient had spondyloarthropathy and fibromyalgia, in addition to her RP. Corticosteroids are the mainstay of treatment but due to steroid side effects, other medications are frequently used (dapson, azathioprine, methotrexate, cyclophosphamide).¹

There are a few reports of RP in the obstetric literature with none mentioning analgesia or anesthesia.⁶⁻⁹ Papo *et al.* reported a 25-year retrospective review of 25 pregnancies that occurred in 11 women after

or concomitantly with the diagnosis of RP.⁸ Disease exacerbations occurred in seven of the pregnancies and five women had other associated inflammatory diseases. Relapsing polychondritis remained asymptomatic during one pregnancy and in 16 pregnancies RP was considered inactive. There were four Cesarean deliveries but the type of anesthetic was not reported. In 26% of women the onset of RP occurred at about the 20th week of pregnancy suggesting a possible hormonal influence.

There are a few reports of the anesthetic management in non-obstetrical patients,¹⁰⁻¹⁴ particularly associated with stenting of the bronchi. These reports highlight the potential problems of airway management including airway obstruction. Airway obstruction may occur due to inflammatory swelling, scar tissue formation and dynamic airway collapse (secondary to progressive destruction of laryngeal, tracheal and bronchial cartilages). Endoscopy and intubation may lead to a marked increase in dyspnea and possibly death, secondary to inflammation and/or airway collapse.⁴ A smaller endotracheal tube may be required for intubation and positive pressure ventilation may be necessary to prevent airway collapse.¹³

The management of the parturient with RP must be multidisciplinary (rheumatology, perinatology, anesthesiology) to ensure optimum care. Establishing the degree of respiratory and cardiovascular involvement is critical as it will help guide the subsequent management. In this particular case pulmonary function studies and flow volume loops prior to pregnancy were normal, as was a bronchoscopy. Although the patient reported palpitations, cardiac dysrhythmias were not identified on a 48-hr Holter monitor evaluation. She did not have a transthoracic echocardiogram as she had no symptoms or physical findings indicative of cardiac involvement.

In order to minimize complications, medical management may have to change as pregnancy progresses, especially when exacerbations occur. This was important with the present case at 33 weeks gestation when hoarseness became severe and there was increased tenderness over her larynx and trachea.

The engorgement and edema of the airway that is normal during pregnancy may worsen respiratory symptoms in RP patients. Dyspnea, a common symptom during normal pregnancy, may indicate airway compromise in RP.¹⁰ The increase in cardiac output and the softening of the media of the vasculature may increase the risk of aneurysmal dilatation of large blood vessels, such as the aorta.

The literature does not resolve a preferred mode of delivery and Cesarean delivery should be based upon

obstetrical considerations. Depending upon the status of the respiratory and cardiovascular systems and presence of concomitant disease, it may be prudent to shorten the second stage of labour through the use of forceps or vacuum. Early provision of epidural analgesia is advisable as analgesia decreases the stress of labour, and ablates the increase in cardiac output during labour. This is of particular importance if there is evidence of vascular or valvular involvement. In the patient with respiratory symptoms, the increased work of breathing and increased oxygen consumption during labour may cause collapse of affected parts of the airway. Effective analgesia should lessen these respiratory effects.

Regional anesthesia is preferred for Cesarean delivery as one wants to avoid airway manipulation. This was particularly true for our patient who had laryngeal symptoms. General anesthesia is problematic for several reasons. As patients with RP have an increased propensity for gastroesophageal reflux, acid aspiration prophylaxis is important. Management of rapid sequence induction and intubation may be difficult as cricoid pressure may be relatively contraindicated if the woman has tracheal tenderness. As noted above, a smaller endotracheal tube may be required in some RP patients because of the inflammatory swelling and scarring of the upper airway. There is little to choose between spinal and epidural anesthesia although an epidural may result in greater hemodynamic stability because of the gradual onset of sympathetic block. This may be important in the woman with valvular or vascular symptoms. Obviously the choice of anesthetic technique will be dependent on the status of each individual patient; the degree of RP involvement (e.g., trachea, aortic or mitral valve, vasculitis) and the obstetric situation (vaginal or operative delivery and urgency of delivery).

In summary, we present a patient with RP in pregnancy. The importance of multidisciplinary cooperation in managing these high risk pregnant patients is emphasized.

References

- 1 *Trentham DE, Le CH.* Relapsing polychondritis (clinical review). *Ann Int Med* 1998; 129: 114–22.
- 2 *Harisdangkul V.* Relapsing polychondritis. In: Klippel JH (Ed.). *Primer on the Rheumatic Diseases*, 12th ed. Atlanta, GA: Arthritis Foundation; 2001: 419–22.
- 3 Appendix 1. Criteria for the classification and diagnosis of the rheumatic diseases. In: Klippel JH (Ed.). *Primer on the Rheumatic Diseases*, 12th ed. Atlanta, GA: Arthritis Foundation; 2001: 631, 635.
- 4 *Eng J, Sabanathan S.* Airway complications in relapsing polychondritis. *Ann Thorac Surg* 1991; 51: 686–92.
- 5 *Selim AGA, Fulford LG, Mohiaddin RH, Sheppard MN.* Active aortitis in relapsing polychondritis. *J Clin Pathol* 2001; 54: 890–2.
- 6 *Bellamy N, Dewar CL.* Relapsing polychondritis in pregnancy. *J Rheumatol* 1990; 17: 1525–6.
- 7 *Gimovsky ML, Nishiyama M.* Relapsing polychondritis in pregnancy: a case report and review. *Am J Obstet Gynecol* 1989; 161: 332–4.
- 8 *Papo T, Wechsler B, Bletry O, Piette AM, Godeau P, Piette JC.* Pregnancy in relapsing polychondritis. Twenty-five pregnancies in eleven patients. *Arthritis Rheum* 1997; 40: 1245–9.
- 9 *Tsanadis GD, Chouliara ST, Voulgari PV, Makrydimas GV, Drosos AA.* Outcome of pregnancy in a patient with relapsing polychondritis and pyoderma gangrenosum (Letter). *Clin Rheumatol* 2002; 21: 538.
- 10 *Burgess FW, Whitlock W, David MJ, Patane PS.* Anesthetic implications of relapsing polychondritis: a case report. *Anesthesiology* 1990; 73: 570–2.
- 11 *Hayward AW, Al-Shaikh B.* Relapsing polychondritis and the anaesthetist. *Anaesthesia* 1988; 43: 573–7.
- 12 *Biro P, Rohling R, Schmid S, Matter C, Lang M.* Anesthesia in a patient with acute respiratory insufficiency due to relapsing polychondritis. *J Clin Anesth* 1994; 6: 59–62.
- 13 *Fitzmaurice BG, Brodsky JB, Kee ST, Foppiano LE, McNutt J.* Anesthetic management of a patient with relapsing polychondritis. *J Cardiothorac Vasc Anesth* 1999; 13: 309–11.
- 14 *Tso AS, Chung HS, Wu CY, et al.* Anesthetic management of a patient with relapsing polychondritis - a case report. *Acta Anaesthesiol Sin* 2001; 39: 189–94.