**Introduction**

Port-wine stains (nevus flammeus, port-wine nevus) are caused by the progressive ectasis of vessels within the cutaneous superficial vascular plexus. They are always present at birth, occur in 0.1% to 0.3% of children, and are not hereditary. Port-wine stains are usually unilateral and most commonly are distributed on the head and neck. In infants, biopsy specimens reveal no vascular wall abnormality, but with advancing age the vessels become progressively dilated and filled with red blood cells. As they become engorged with blood, port-wine stains darken to the characteristic purple hue. In approximately two-thirds of patients, the tissues in the affected area may hypertrophy. The mechanism causing this abnormality is thought to be altered neural regulation of the vasculature in the area of the port-wine stain. The tissue surrounding the stain is at greater risk for engorgement or edema with any alteration in blood flow or changes in the Starling dynamics within the vascular space. The relative position of the port-wine lesion in relationship to a patient's position for surgery and the type and amount of intravenous (IV) fluids administered to the patient also can have an impact on the potential for edema or tissue engorgement in the area of the lesion. Finally, anesthetic agents with vasoactive properties also may affect the management of patients with this lesion because port-wine stains have been reported to fade during the use of inhalation anesthetic agents, making the abnormality more difficult to detect under anesthesia.

Patients with port-wine stain have powerful psychological responses to their lesions. Coping mechanisms are developed as a result of a variety of very strong emotional reactions, including a profound sense of embarrassment. Patients with prominent lesions are compelled to conceal the port-wine stain in public and in the presence of family members. With the development of effective, long-lasting, waterproof cover-all makeup, patients with very significant port-wine stains on a visible body part may progress through the busy, time-pressured healthcare system with the stain undetected. As demonstrated in this case report, the psychological component of this patient's large facial port-wine stain played a key role in the evolution of perioperative events that placed the patient at risk for a adverse outcome.

**Case report**

A 63-year-old woman arrived at the surgical suite of a large, tertiary medical center for the purpose of donating her left kidney to her 28-year-old daughter, who had end-stage renal disease. The preanesthetic interview took place in the preoperative holding area with the daughter present on the adjacent gurney. The patient described herself as being in excellent health with good exercise tolerance and no history of smoking or alcohol use. Physical examination revealed a healthy woman appearing younger than her stated age, (Figure, View A). A large left elbow hematoma was noted, and the patient stated that it was a result of a recent fall. The patient's medical history included diet-controlled hypertension. Her surgical history included an exploratory laparotomy with no anesthesia-related complications.
An 18-gauge IV catheter was placed in the patient’s right anterior forearm for a 0.9% normal saline solution (NSS) infusion. Midazolam, 1 mg IV, was administered for preoperative sedation, and 500 mL of the infusing NSS was given rapidly at the request of the surgeon. The patient was transported to the operating room, monitors were placed, and induction of anesthesia was performed without complication. Induction agents included fentanyl, 150 µg; curare, 3 mg; thiopental, 300 mg; and succinylcholine, 120 mg. The patient was orally intubated with a 7.0 mm Sheridan endotracheal tube (Kendall Co, Mansfield, Mass), and a 39% oxygen, 60% nitrous oxide, 1% isoflurane inhalation mixture was initiated. During the taping of the endotracheal tube, it was noted that the patient was wearing makeup. An additional 16-gauge IV catheter was started in the left anterior forearm, and a right radial arterial line was placed.

The patient was placed in a right lateral decubitus position for the left nephrectomy surgery. The surgeon requested flexion of the table to enhance operative access to the left kidney. As a result, the patient’s head rested in a slightly dependent position in relation to the rest of her torso. An axillary roll was inserted, bony prominences were padded, and the right eye and ear were within the open space of a foam “donut-style” head rest. Maintenance medications included oxygen, nitrous oxide, isoflurane, and incremental fentanyl (2 µg/kg per hour). Muscle relaxation was maintained with cisatracurium. The patient was ventilated to maintain normocapnia as determined by end-tidal carbon dioxide monitoring. Furosemide and mannitol were given after an intentional mild hypervolemic state was created with the administration of NSS. Systolic blood pressure was maintained between 130 to 140 mm Hg per surgeon request. Following the diuretic administration, urine output ranged from 80 to 140 mL every 15 minutes. The left kidney was removed without complication. The right eye and ear remained within the opening of the donut as evaluated and documented every 15 minutes. The procedural duration was 3.5 hours, and fluid totals included 5,500 mL of IV NSS, 1,400 mL of urinary output, and an estimated blood loss of 100 mL. As the dressing was placed on the left flank, neostigmine, 3 mg, and glycopyrrolate, 0.6 mg, were given to reverse the neuromuscular blockade.

The patient was then turned to a supine position. Upon turning of the patient, a pronounced right-sided facial edema and a large, question-mark shaped purple discoloration extending from the right temple to the right mandible were immediately noted (Figure, View B). The most significant area of edema was in the periorbital region and approximated the shape and size of the “hole” in the foam donut positioning device. When palpated, the edema was soft and nonpitting, and
underlying structures were easily identified. The right eye was swollen shut and could not be examined. Before the patient was fully awake, careful intraoral visualization revealed no airway edema or abnormal anatomy.

After discussion, the decision was reached to extubate the patient when she met extubation criteria. Soon after, the patient was extubated without complications. She was transported in stable condition with oxygen via face mask to the postanesthesia care unit (PACU). Upon arrival in the PACU her vital signs were stable, and the patient was placed in a semi-Fowler’s position. The skin over the discolored, edematous area was warm, and the purple discoloration became even more vivid when the right facial area was palpated. The patient was unable to open her right eye but did not express discomfort with examination of the area. A warm compress was applied to the right facial area after report was given to the PACU nurse. Approximately 90 minutes later, the patient was reevaluated. The right-sided edema had receded dramatically, and the patient could open her right eye normally. Extraocular movements and a vision examination of the right eye were performed with no deficits noted. The right side of the patient’s face was palpated and was warm to touch; however, the large purple discoloration remained (Figure, View C).

When queried, the patient reported no discomfort and noted that at least 5 other providers had been in to evaluate her face on that side, but no one had yet told her if her daughter’s surgery had gone well. Her daughter was across the room, and her new kidney was making clear yellow urine. The primary author relayed this to the patient, who began to cry. The author then explained to the patient that the entire right side of her face had been very swollen with a large purple discoloration area of unknown etiology. The patient asked, “Is it different from the port-wine stain birthmark on my face?” Stunned, the author then asked the patient why she had not revealed this large vascular abnormality in the preanesthesia interview. The patient stated that her 28-year-old daughter, who had been lying on the adjacent gurney cart during the preoperative interview, had never seen the lesion. Upon further questioning, the patient indicated that she had carefully kept her children unaware of this birthmark by using makeup and avoiding situations that would reveal the lesion to her family, eg, swimming and exercise. She then asked that her makeup kit be brought to the PACU so that she could reapply her cover-all foundation.

Discussion

• Etiology of the facial edema. Port-wine stains are congenitally acquired and result from abnormal regulation of blood flow that leads to a progressive vascular dilatation and the characteristic discoloration. Research has documented a decrease in perivascular nerve elements around port-wine stains. This deficit may be responsible for altered neural modulation of vascular tone, leading to the progressive vascular changes that characterize port-wine stains. Because general anesthesia alters both neuronal outflow and vascular regulation, patients with port-wine stain lesions may be at risk for additional perioperative changes in their lesions. We speculate that the edema noted in this case may be attributed to several factors, including an altered neurovascular control, an increase in the hydrostatic pressure within the capillary bed of the lesion due to the dependent position of the patient’s head, the large volume of crystalloid infused, and maintenance of a high-normal blood pressure. All of these factors have an impact on Starling forces, which govern the maintenance of intravascular volume. According to the Starling principles that govern fluid movement within the capillary bed, a combination of increased hydrostatic pressure (dependent position and elevated blood pressure) and decreased colloidal osmotic pressure (normal saline dilution of vascular volume) would work to increase net fluid movement out of the capillary and into the interstitial space. We speculate that this effect was amplified because of the altered vascularity in the area of the port-wine stain.

• Implications of the facial/periorbital edema. The facial and periorbital edema that occurred in this case was alarming. The patient’s soft tissue swelling was pronounced (Figure, View B) and prevented evaluation of eye function. Before her disclosure of the port-wine stain, we were concerned that the edema and discoloration were evidence of a positioning-related injury involving the eye. Our greatest fear was that the patient would suffer vision loss in the postoperative period.

Ophthalmic venous obstruction is a relatively uncommon positioning-related complication that can lead to acute postoperative vision loss. The etiology is external compression of the eye that obstructs venous drainage and raises intraocular pressure. Perfusion pressure to the optic nerve is the difference between arterial and venous pressure, or between arterial and intraocular pressure if intraocular exceeds venous pressure. Should intraocular pressure exceed venous pressure, retinal artery hemorrhage may occur, and when intraocular pressure exceeds retinal artery pressure, retinal and optic nerve ischemia is possible.

Acute postoperative vision loss following general
Anesthesia is most commonly associated with a condition referred to as ischemic optic neuropathy. This condition is subcategorized into anterior ischemic optic neuropathy and posterior ischemic optic neuropathy with respect to the vascular supply involved and the visual fields affected. Infarction of the optic nerve occurs as a result of inadequate oxygen delivery from small feeder arterioles. Ischemic optic neuropathy also was a possibility in this patient because inadequate optic nerve oxygenation can occur in the setting of increased intraocular pressure or venous engorgement, leading to increased resistance to arterial blood flow.

- Inhalation anesthetic agents and port-wine stain. Why did we fail to detect the purple discoloration during the procedure? We speculate that the cover-all makeup was removed through contact of the right side of the face with the foam head rest and the linens during the positioning of the patient. The view of the right facial area was obscured by the right lateral decubitus position and positioning of the head in the donut. In addition, isoflurane was used for maintenance during the case. Isoflurane and sevoflurane have been reported to cause port-wine stain lesions to fade or disappear, making the lesions difficult to detect. This fading has been attributed to decreased perfusion through an increase in the arterial blood flow in the abnormal port-wine stain vasculature due to vasodilation of blood vessels peripheral to the lesion. The effect is analogous to the “steal phenomenon,” which has been widely discussed in both cardiac and neuroanesthesia textbooks. Theoretically, steal occurs when vessels surrounding regions with abnormal vasculature dilate. Blood is thus diverted from the region of abnormal vasculature to surrounding tissues.

- Psychology of patients with a port-wine stain. While the natural progression of port-wine stains may lead to medical complications, most research indicates that the major morbidity of port-wine stain is psychological. The value our society places on facial appearance is high. The negative psychological impact of a port-wine stain occurs throughout a person’s lifetime. Patients with port-wine stains may experience feelings of stigmatization, embarrassment, anxiety, and depression. In an analysis of the psychological stress that ensues with port-wine stains, patients showed personality changes when removing cosmetic camouflage by becoming withdrawn and avoiding eye contact. The amount of personal embarrassment is not necessarily restricted to women, although more than 50% of female patients will not appear in public without makeup, and a full third of female patients would not show their birthmark to their spouse or children. The patient described in this case study was displaying behavior that is consistent within this population. She was compelled to keep her makeup on due to fear of public disclosure and the presence of her daughter on the adjacent gurney. Because the anesthesia providers were unaware of the facial port-wine stain during the perioperative period, the patient was placed at risk for the development of lesion-related complications.

- Anesthetic approaches if lesion had been disclosed. Had we been aware of the lesion preoperatively, several changes in the intraoperative management of the patient could have been considered. First, an alternative surgical approach with an anterior abdominal incision would have allowed the patient to remain supine. If the surgeon indicated that the lateral flank incision was absolutely necessary, then the degree of head-down angle in the lateral, flexed position could have been minimized or eliminated.

A second consideration in the anesthetic management would have involved the type and amount of IV fluids infused to the patient. For patients undergoing living-related donor kidney resection, renal perfusion is critical. Routinely, the choice of fluids is a balanced salt solution (0.9% NSS) that is used to promote renal perfusion through an increase in the arterial blood pressure. For this case, judicious colloid administration might have been an acceptable substitute for the NSS. Due to the superior ability of colloid solution to remain in the intravascular space while still achieving the goal of elevated renal perfusion pressure, we speculate that the patient’s facial edema could have been minimized with judicious colloid administration.

**Summary**

Anesthesia implications for a patient with a port-wine stain include careful choice of surgical position, frequent intraoperative assessment and documentation of the lesion’s status, understanding of the interaction between inhalation agents and the lesion, and the need to be sensitive to the profound psychological impact of the lesions. Should surgical need require dependent position of the port-wine stain, careful padding and frequent evaluation of the stain, with scrupulous documentation of these activities, are indicated. Careful selection of fluid amount and type, as well as frequent assessment of the affected area for edema or change in appearance, are key interventions that may prevent the development of intraoperative complications associated with port-wine stains.

**REFERENCES**


AUTHORS
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