Treatment of Congenital Subglottic Hemangiomas

Our Experience Compared With Reports in the Literature

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Objective: To evaluate the outcome of our experience in the treatment of congenital subglottic hemangiomas.

Design: Retrospective review of records.

Setting: Airway tertiary care service.

Patients: From 1986 to 2006 we treated 39 pediatric patients affected by congenital subglottic hemangiomas.

Intervention: Therapeutic choice depended on presentation symptoms and grade of respiratory obstruction: 6 patients were primarily treated with only systemic steroids; 11 patients underwent intralesional corticosteroid injections followed by tracheal intubation and systemic steroid support; and 22 patients underwent primary diode laser treatment.

Main Outcome Measure: The outcomes were evaluated according to 1 or more of the following criteria: resolution of symptoms, reduction of airway obstruction, the need and duration of intubation, tracheotomy decannulation, need of further treatments, and occurrence of complications.

Results: Patients treated with only systemic steroids showed a success rate of 50% (3 of 6); patients who underwent intralesional corticosteroid injections followed by tracheal intubation and systemic steroid support reached a positive result in 73% of cases (8 of 11). On the whole, 18% of patients treated with full-dose systemic steroids developed significant adverse effects (3 of 17). The success rate was 95% among patients treated with diode laser as primary treatment (21 of 22), with a complication rate of 9% (2 of 22).

Conclusions: Endoscopic laser surgery is the therapeutic option that most approaches the objectives of securing the airway while using the least invasive method possible and reducing to a minimum the necessity and duration of intubation. On the basis of our experience, we believe that diode laser, owing to its physical and structural features, is the safest and most effective device for the treatment of congenital subglottic hemangiomas. Treatment with intralesional or systemic corticosteroids could have an adjuvant role.


VEN THOUGH HEMANGIOMAS ARE THE MOST COMMON HEAD AND NECK TUMORS IN PEDIATRIC PATIENTS, THEIR SUBGLOTTIC LOCALIZATION IS QUITE RARE, ACCOUNTING FOR ONLY 1.5% OF ALL CONGENITAL LARYNGEAL LESIONS.1 THE SYMPTOMS OF THIS DISEASE ARE STRICTLY RELATED TO THE GRADE OF AIRWAY OBSTRUCTION, WHICH BECOMES MORE EVIDENT DURING PERIODS OF AGITATION, CRYING, OR RESPIRATORY INFECTIONS. STRIDOR, USUALLY BIPHASIC BUT MORE PROMINENT DURING INSPIRATION, IS THE MOST COMMON PRESENTATION SYMPTOM OF CONGENITAL SUBGLOTTIC HEMANGIOMAS (CSGH). IN THE CASES OF WIDER LESIONS, THE SIGNS OF RESPIRATORY DISTRESS ARE PRESENT WITH BARKING COUGH, THORACIC AND ABDOMINAL RECESSIONS, HOARSENESS, AND CYANOSIS.2 USUALLY THESE SYMPTOMS ARE NOT PRESENT AT BIRTH BUT ARISE IN THE FIRST 3 MONTHS OF LIFE AFTER A RAPID PROLIFERATIVE PHASE THAT PEAKS AROUND AGE 1 YEAR AND THEN STARTS A GRADUAL INVOLUTIONAL PROCESS LEADING TO COMPLETE RESOLUTION OVER A 2- TO 5-YEAR PERIOD.3 THE SELF-LIMITING COURSE OF THIS DISEASE MUST BE KEPT IN MIND WHEN CHOOSING A TREATMENT. A VARIETY OF THERAPEUTIC OPTIONS HAS BEEN PROPOSED IN PREVIOUS YEARS TO TREAT CSGH, INCLUDING EXTERNAL IRRADIATION,4 TRACHEOTOMY,5 SURGICAL EXCISION,6 SYSTEMIC OR INTRALESIONAL CORTICOSTEROIDS,7-9 LASER VAPORIZATION,9 AND INTERFERON.10

The aim of this work was to analyze our experience with endoscopic treatment of CSGH and to compare our data with those reported in literature.

METHODS

We carried out a retrospective medical records review of all patients treated for CSGH in our institution during a period of 20 years (1986 to 2006). For each patient, the following char-
acteristics were considered: sex, age at first diagnosis, signs and symptoms, diagnostic assessment, site of the tumor, grade of airway obstruction, and treatment techniques.

In all cases, after the routine assessment (clinical evaluation, biochemical examinations, and chest radiography), the diagnosis was made by means of an endoscopic direct visualization of the tumor under general anesthesia (Figure 1). We used rigid rod-lens telescopes with diameters of 2.7 and 4 mm with either direct or 30° angled vision.

Therapeutic choice depended on symptoms and grade of respiratory impairment. Patients presenting with only inspiratory stridor and airway obstruction up to 70% (group 1) were treated primarily with oral steroids and monitored until the symptoms resolved. We administered dexamethasone sodium phosphate (1 mg/kg/d) for 7 days followed by a slow decrease of the doses every 48 to 72 hours until reaching minimum levels. Patients with signs of moderate to severe respiratory distress and/or airway obstruction greater than 70% (group 2) were treated with 2 different approaches. In the first 10 years of our experience (group 2A), we administered an intraleisional corticosteroid injection followed by intubation and treatment with systemic dexamethasone. Since 1995, we treated hemangiomas with laser vaporization (Figure 2) using a diode laser (Biolitec AG, Jena, Germany) under videoendoscopic control (group 2B).

To reduce the variability due to cumulative effects of different subsequent treatments on each patient, only the primary treatments were considered in the present analysis. The outcomes of these treatments were evaluated according to 1 or more of the following criteria: resolution of symptoms, reduction of airway obstruction, the need and duration of intubation, tracheotomy decannulation, need of further treatments, and occurrence of complications.

All the data revealed by our work were then compared with the main experiences reported in the English-language literature.

RESULTS

From 1986 to 2006, 39 pediatric patients affected by CSGH were treated at the Division of Airway Endoscopic Surgery, Padua University Hospital. There were 30 girls and 9 boys (ratio, 3.3:1) with a mean age at the first diagnosis of 5.1 months (range, 1.3-15.7 months). All patients were referred to us for respiratory problems: stridor was present in 13, while 26 patients showed signs and symptoms of moderate to severe respiratory distress. In 20 cases, the lesion was localized in the left portion of the subglottic region, while in 8 patients, the right side was involved. In 8 cases, the hemangioma was posterior, and in 2 patients it was bilateral.

Six patients (group 1) were primarily treated with only systemic steroids for a mean period of 6.4 weeks (range, 3 weeks to 6 months); 3 of them achieved stable control of symptoms, while 1 of them experienced a recurrence of stridor after therapy suspension that required diode laser vaporization. The same secondary laser treatment was used in 2 cases in which a progression of the tumor with worsening of respiratory symptoms was recorded notwithstanding steroid treatment.

Eleven patients (group 2A) underwent intraleisional corticosteroid injection followed by tracheal intubation and systemic steroid support. The mean intubation period was 8.3 days (range, 5.0-15.0 days). In 3 of 11 patients, a tracheotomy was carried out owing to inability to remove the ventilation tube after 15 consecutive days; 2 of these 3 patients underwent secondary laser treatment. One patient who was referred to us had already undergone a tracheotomy. The mean decannulation time was 14.7 months (range, 13.1-17.4 months).

In 22 patients (group 2B), diode laser was used as the primary treatment of CSGH, with a mean of 1.6 sessions (range, 1.0-3.0) performed for each patient at a mean interval of 3.5 months (range, 1.3-13.4 months). In 6 of 22 cases, laser vaporization was followed by tracheal intubation (27%) for a mean period of 3.7 days (range, 2.0-5.0 days). In 21 of 22 cases (95%), remission of symptoms and a significant reduction of the tumor’s dimensions was obtained (mean reduction, 68%; range of reduction, 55%-100%) as shown by endoscopic control. One patient, affected by moderate gastroesophageal reflux disease, underwent a tracheotomy owing to the persistence of respiratory symptoms after 3 laser sessions. In...
2 cases, a tracheotomy was performed preventively before laser treatment for associated diseases (1 case of severe gastroesophageal reflux disease that required Nissen fundoplication and 1 case of cardiovascular dysfunction). Two other patients who were referred to us had already undergone a tracheotomy. The mean decannulation time was 11.3 months (range, 9.0-14.0 months).

As for complications, 3 of the 17 patients treated with full-dose systemic steroids (groups 1 and 2A) developed adverse effects (18%), which were controlled with a decrease of steroid dose in 2 cases (hypertension and gastropathy) and suspension of treatment in 1 (a patient with Cushing disease who was subsequently treated with laser treatment). No complications strictly caused by intralesional corticosteroid injections (group 2A) were observed in any patients. No early complications (in particular, intraoperative or postoperative bleeding) were verified in the 22 patients who underwent laser surgery (group 2B); in 2 cases minor late complications that required further treatment occurred (9%): a tracheal posterolateral bridge synechia (resolved after treatment with a diode laser section) and a laser-induced subglottic granuloma (successfully removed by means of videoendoscopic forces). No secondary subglottic stenoses were recorded.

COMMENT

Although it is clear that the aim of any treatment is to secure the airway, the rarity of CSGH and the consequent scarcity and variability of any single experience give rise to a lack of consensus and unequivocal guidelines to direct treatment of this disease. To provide clearer direction, Bitar et al reviewed all the cases of CSGH reported in the English-language literature between 1986 and 2002 (28 series including 372 patients over the 17-year period). From this work, it appears that carbon dioxide laser vaporization was the most widely used technique for treatment of CSGH (181 of 372), followed by tracheotomy (164 of 372), systemic corticosteroids (163 of 372), intralesional steroids (54 of 372), and open surgical excision (50 of 372). Other techniques are only occasionally used.

In 81 patients, carbon dioxide laser vaporization was used as the primary treatment, with a success rate of 88.9%. Patients underwent a mean of 2 treatment sessions (range, 1-5 sessions), and a complication rate of 12.2% was reported, in particular subglottic stenoses (5.5%). In our experience with a smaller number of patients, diode laser was the primary treatment, showing a success rate of 95% (21 of 22) and an incidence of minor complications of 9% (2 of 22) without either early or late major complications.

Several considerations lead us to believe that diode laser therapy is particularly suitable and effective in the treatment of CSGH. First, the diode laser’s beam, in contrast to the carbon dioxide laser beam, is carried by flexible optic fibers that can be coupled with telescopes permitting access to sites such as the subglottic region that are difficult to explore with other techniques. Moreover, the exclusive use of this laser by contact or extremely close distance makes it much safer than other laser sources by avoiding damage due to “beam escape” in an open field. Other advantages are related to the physical features of the diode laser: its specific wavelength (810-980 nm) is not only absorbed by water (although less so than carbon dioxide laser wavelength) but also by other chromophores such as melanin and, in particular, oxyhemoglobin. From a practical point of view, all of these features confer on the diode laser a surgical precision only slightly inferior to that of the carbon dioxide laser and a superior capability of coagulation and thermal vaporization, making the diode laser particularly suitable to treat vascular lesions such as CSGH. Diode laser also appears to be more precise and less invasive than other devices such as Neodymium:YAG and potassium-titanyl-phosphate lasers, which have been used to treat hemangiomas in a small number of cases.

The same technical precautions reported for other laser sources also apply to diode laser surgery. In particular, to prevent the evolution of scarring, damage to the perichondrium must be avoided. Similarly, excessively long and frequent laser exposure must be avoided. In our experience, we find it appropriate to submit the patient to no more than 3 laser sessions, with a minimum interval of 1 month between sessions. To favor vaporization over than section, we use the diode laser at a medium power emission (5-8 W) in continuous mode or with low-frequency pulsation, and with fibers of 300 to 600 μm. We also reduce irradiation time to a minimum to avoid thermal damage as much as possible. For the same reason, we cool the surgical field with a cotton sponge soaked with isotonic sodium chloride solution at 4°C.

With regard to corticosteroid treatment, Bitar et al reported that intrallesional steroid injection, used as primary treatment for 22 of 372 patients, showed a success rate of 86.4% with a mean of 3 injections. However, this treatment required a mean intubation period of 38 days. In our opinion, such a long intubation period exposes the patient to an unacceptable risk of early or late airway injuries for the treatment of a self-limiting disease. For this reason, we performed a temporary tracheotomy on all patients we could not wean from the ventilation tube after a 15 days (4 of 11 patients who were treated with steroids and 1 of 22 patients who underwent diode laser treatment). The rather high incidence of tracheotomy in patients treated with intrallesional steroids is owing to the long action time associated with this treatment, in contrast to the immediate effect of laser vaporization.

The use of systemic steroids, although widely reported in literature as the primary treatment (102 of 372), showed a poor success rate, particularly for large tumors. Moreover, this therapy is associated with adverse effects (Cushing disease, growth retardation, hypertension, and immunodeficiency) reported in 12.9% of the cases and verified in our experience. On the other hand, we believe that systemic steroids, administered at controlled doses and for a short period, could have an adjuvant role after laser treatment.

Open surgical excision of CSGH, first described by Sharp in 1945 and recently revisited by others, showed a success rate of 98%. It was used on 50 of 372 patients, re-
quired a mean intubation or stenting period of 9 days, and carried a 10% complication rate, complications including subglottic stenosis, bleeding, and wound infections.\textsuperscript{12} We believe that this technique, while efficacious, is too invasive as a primary procedure and therefore should be considered only for selected cases not suitable for endoscopic surgery or after failure of other treatments.

The main goal of any CSGH treatment is to secure the airway as conservatively as possible and reduce to a minimum the intubation period. Considering the pathologic features and the clinical evolution of CSGH, we believe that endoscopic laser surgery is the therapeutic option that most effectively achieves these objectives. We also believe that diode laser, owing to its physical and structural features, is the most effective and safe device in the treatment of CSGH. The use of intralesional or systemic corticosteroids could have a helpful adjuvant role to laser treatment.

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Author Contributions: Dr Saetti had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Saetti and Narne. Acquisition of data: Saetti, Silvestrini, and Cutrone. Analysis and interpretation of data: Saetti and Silvestrini. Drafting of the manuscript: Saetti and Cutrone. Critical revision of the manuscript for important intellectual content: Saetti, Silvestrini, and Narne. Administrative, technical, and material support: Saetti, Silvestrini, and Cutrone. Study supervision: Saetti and Narne.

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