Posttransplant Lymphoproliferative Disorder involving the Larynx

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ABSTRACT

Aim: Our goal is to present an unusual case of posttransplant lymphoproliferative disorder (PTLD) involving the larynx. We aim to expand the literature regarding head and neck manifestations involving PTLD and for otolaryngologists to include PTLD in their differential diagnosis of laryngeal lesions in patients who have a history of solid organ transplantation.

Background: Posttransplant lymphoproliferative disorder occurs in immunocompromised patients following solid organ transplantation. Head and neck manifestations most commonly involve Waldeyer’s ring with the larynx and trachea being relatively uncommon sites of disease. However, lesions of the larynx can cause acute airway obstruction and rare fatalities have been reported in the literature.

Case report: We present the case of a 51-year-old female with a history of renal transplantation, who presented to the office after an incidental supraglottic lesion was discovered during intubation for cochlear implant placement. A mucosalized lesion arising from the right aryepiglottic fold with dynamic obstruction of the airway was noted on office endoscopic examination. Imaging revealed extension of the mass into the right posterolateral hypopharynx. The patient was taken to the operating room for microdirect laryngoscopy and CO2 laser excision of the mass. The lesion was excised with negative margins. Final pathology revealed plasmacytoma-like lesion in a posttransplant patient. Special emphasis should be given to the tissues of Waldeyer’s ring and larynx.

Conclusion: We present the case of an incidental supraglottic lesion in a posttransplant patient. Special emphasis should be given to the tissues of Waldeyer’s ring and larynx.

Clinical significance: Posttransplant lymphoproliferative disorder, while rare, should be kept in the differential diagnosis of pharyngeal and airway lesions in patients with a history of solid organ transplantation. Expansile lesions can cause acute airway obstruction. Urgent evaluation and treatment of these lesions is necessary in these circumstances.

Keywords: Adult, Airway obstruction, Immunocompromised, Laryngology, Posttransplant lymphoproliferative disorder.

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Demographically, children are most commonly affected with a threefold increased chance of developing PTLD thought to be secondary to the higher incidence of EBV in the pediatric population. A retrospective case series by Lattyak et al estimated that approximately 10% of pediatric patients undergoing liver transplantation will eventually develop PTLD in the head and neck.

Patients with laryngeal or tracheal lesions may present with signs of acute airway obstruction, such as stridor, cough, and dyspnea. Other times, clinical findings may resemble the signs of infectious mononucleosis including fevers, malaise, odynophagia, and lymphadenopathy. On physical exam, hypertrophy of the tissues of Waldeyer’s ring may be noted in addition to mucosal masses of the larynx and trachea on laryngoscopy.

A case series of children undergoing liver transplantation was reviewed at a Belgian hospital in a 2002 prospective study. In this study, 77 pediatric liver transplant recipients were followed for a 3-year period and monitored for the development of PTLD in the head and neck. Eight of these patients developed acute nonbacterial tonsillitis and those who were positive for EBV (as noted by positive IgM antibodies) underwent tonsillectomy. These patients also underwent concomitant reduction in the level of immunosuppression.

While the larynx and trachea appear to be less common sites of PTLD, multiple case reports of PTLD have been reported involving the upper airway. For example, a 6-year old boy with a history of liver transplantation presented to the hospital with stridor and was taken to the operating room for emergent direct laryngoscopy, bronchoscopy and tracheostomy. On bronchoscopy, a 1 to 2 cm subglottic granulomatous mass was found causing over 90% airway obstruction.

One episode of sudden respiratory arrest and death secondary to PTLD was found in the literature. A 3-year old female on cytotoxic therapy following heart transplantation presented with persistent stridor despite treatment for suspected viral laryngotracheitis. Following admission, the patient experienced acute airway obstruction and was subsequently underwent renal transplantation who presented with stridor and was noted to have edema of the epiglottis. While laryngeal biopsy did not reveal the source of the lesion, PTLD was confirmed following tonsillectomy and adenoidectomy.

Banks et al reported on a case of PTLD involving the larynx and trachea. The patient had a history of congenital nephrotic syndrome, bilateral nephrectomies, and subsequently underwent renal transplant at 10 months of age. The patient had previously presented with cervical lymphadenopathy that was positive for PTLD on open biopsy. Several years later, the patient presented to the emergency room with fevers, worsening cough, and shortness of breath with computed tomography (CT) chest revealing a paratracheal mass. The patient then underwent direct laryngoscopy, bronchoscopy, and biopsy of the tracheal mass as well as a soft tissue mass of the right laryngeal ventricle that was positive for PTLD. The patient’s lesions eventually resolved following the reduction of immunosuppressive therapy and initiation of chemotherapy.

Lesions of the oral cavity, while rare, have also been reported in the literature. A 2008 case series reported 3 cases of PTLD in the oral cavity following organ transplantation. These lesions involved the oral tongue, gingiva and buccal vestibule. These lesions involved the buccal mucosa and gingiva and were treated with wide local excision and modification of immunosuppression.

While no randomized control trials or guidelines have been published to delineate the standards of care for management of PTLD, treatment of the disease involves reduction of the immunosuppressive agent by up to 50%. However, there is a risk of graft rejection in these cases thus monitoring of graft function is necessary. Further, patients require close monitoring for clinical deterioration and weekly EBV evaluation may be performed to monitor viral load. Cases refractory to this treatment may require antivirals, chemotherapy, immunotherapy, or definitive surgical resection of the offending lesion. In some cases, positron emission tomography and computed tomography (PET-CT) may be used for posttreatment surveillance to evaluate for resolution of the disease.

CASE REPORT

The patient is a 51-year-old woman with a past medical history of renal failure and renal transplant 18 years prior to presentation. She was referred for laryngology evaluation after attempted intubation for elective cochlear implantation secondary to profound sensorineural hearing loss. During laryngoscopy, intubation was aborted when the anesthesiologist encountered an obstructing supraglottic mass.
The patient denied any shortness of breath or difficulty swallowing. She had a normal voice, no stridor, and denied globus sensation. Flexible laryngoscopic examination revealed a solid appearing, smoothly mucosalized mass involving the right aryepiglottic fold (Fig. 1). This mass appeared to be dynamically obstructing the airway. However, the glottic and subglottic airway was normal. Computed tomography scan of the neck with intravenous contrast showed an enhancing, solid mass, which was contiguous with the right aryepiglottic fold. There were no cystic components seen and the lesion protruded into the right posterolateral hypopharynx (Figs 2 and 3).

The patient was taken to the operating for microlaryngoscopy with CO₂ laser resection of the mass. After communicating with the anesthesiologist, the patient was safely intubated. The lesion was resected with appropriate gross margins. After removal, the airway was clear and widely patent. The patient was extubated without difficulty. Pathology revealed plasma cells consistent with Plasmacytoma-like PTLD (Figs 4A and B). The patient did well postoperatively and was referred back to her transplant team for reduction of her immunosuppressive regimen.

DISCUSSION

The case above demonstrates an incidental lesion of PTLD involving the supraglottic larynx. While the larynx is an uncommon site for PTLD, multiple case reports of PTLD involving the upper airway have been reported. For patient with a history of solid organ transplantation and presenting symptoms consistent with upper airway obstruction, there must be a high index of suspicion for PTLD. Tissue biopsy is required for the diagnosis. Treatment typically involves reduction of the immunosuppressive agent as well as chemotherapy for non-responders. Antivirals including acyclovir have been used in certain patients as well, which are designed to target EBV.

CONCLUSION

We present on a relatively rare instance of PTLD involving the supraglottic larynx in a patient who underwent bilateral renal transplantation. Posttransplant lymphoproliferative disorder and its head and neck manifestations should be kept in mind when evaluating a patient with history of solid organ transplantation. In particular, additional emphasis on the tissues of Waldeyer’s ring as well as the larynx should be placed to assess for tissue hypertrophy or masses. Surgical biopsy of suspicious or symptomatic lesions should be performed.

CLINICAL SIGNIFICANCE

In this case report, we report on a relatively rare occurrence of PTLD involving the larynx. While this lesion was incidentally discovered, other cases of PTLD involving the larynx and trachea and may be found in patients.
presenting with signs and symptoms of upper airway obstruction. Otolaryngologists should maintain PTLD in their differential diagnosis when managing patients who have a history of organ transplantation. Multiple case reports have highlighted the fact that although rare, PTLD of the larynx and trachea can cause life-threatening airway obstruction. For children developing new onset sleep apnea with adenotonsillar hypertrophy after solid organ transplantation, prompt tonsillectomy and adenoidectomy should be considered as a primary form of management as severe hypertrophy of these tissues may cause acute, life-threatening airway obstruction. The primary role of the otolaryngologist remains for management of the airway, biopsy and debulking of obstructive lesions.

Following surgical management, reduction of immunosuppression is necessary, which requires coordination with the oncologist and transplant team. The addition of antiviral therapy may also be necessary to reduce the EBV viral load. Thus, close communication and coordination is necessary between the otolaryngologist, oncologist, and transplant physicians for optimal management.

REFERENCES


