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Tracheal Chondrosarcoma Excised With Extracorporeal Circulation

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Introduction: Tracheal chondrosarcoma (TCS) is a rare malignancy, with only 19 cases described in the literature to date.

Case presentation: Herein, we presented the third-largest TCS with such an airway compromise that neither orotracheal intubation nor jet ventilation or even tracheostomy was possible. So, extracorporeal circulation was needed to excise the tumor in a one-stage procedure. The patient presented no tumor recurrences after surgery during an approximate7-years follow-up. So open surgical resection and end-to-end anastomosis probes may be applied as a safe and successful treatment. A review of the previous literature revealed no extracorporeal circulation in previous practices.

Conclusion: Management of tracheal chondrosarcoma is challenging due to its airway compromise during the procedure. Different treatment modalities have been advised but none of them included extracorporeal circulation as an option. We believe that this approach allows for better control of the resection and ensures better oxygenation of the patient.

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INTRODUCTION

Tracheal malignancies are not common neoplasms and most of them (50%) are primarily originated in this structure [1]. They may also present secondary to invasion from adjacent structures or as a metastatic mass from other locations. Primary tracheal neoplasms account for 0.2% of all malignant masses in the respiratory tract; being much more frequent than lung or laryngeal neoplasms. Most of these neoplasms (80%) are squamous cell carcinoma and adenoid cystic carcinoma. Carcinoid tumor, adenocarcinoma, and small cell carcinoma remain as the second most common tracheal neoplasms with a much lower incidence [2]. Tracheal chondrosarcomas are an extremely rare malignancy with only 18 cases described in English literature to date [3, 4]. We describe the only case of a patient with obstructive tracheal chondrosarcoma impeding tracheal intubation that required extracorporeal circulation to achieve its complete resection successfully.

CASE PRESENTATION

A 75-year-old male, with a smoking history of 90 pack per year, was presented with progressive dyspnea and stridor for approximately 1 year. He was diagnosed with obstructive sleep apnea syndrome that needed continuous positive airway pressure (CPAP) use. In his past medical history,

he had arterial hypertension, atrial fibrillation, hyperuricemia with secondary arthritis, and renal insufficiency. He was presented with episodes of acute dyspnea for which he received treatment with bronchodilators as a misdiagnosis of chronic obstructive pulmonary disease. He was referred to our department once the pulmonologist observed a tracheal mass that obstructed almost the whole tracheal lumen. The patient had a tracheal mass obliterating the tracheal lumen which impeded the distal pass of the bronchoscope (Figure 1). No biopsy was performed during the procedure. CT scan showed a left side dependent tracheal wall mass of 33x30x38 mm3; obstructing the tracheal airway that left a tracheal lumen of 2mm (Figure 2). The tumor origin was 2 cm below the vocal folds and it presented multiple calcifications within it. Surgery was planned and since it was impossible to perform a tracheostomy underneath the tumor level due to the size of the mass and there was no space left in the trachea to insert neither an endotracheal tube nor jet ventilation in a secure way, extracorporeal circulation was prepared to warranty the oxygenation of the patient during the surgery. An anterior cervicotomy was performed and the larynx, the thyroid isthmus, and the 4 first tracheal rings in which the tumor was confined were exposed. About 4 cm of the trachea; containing the tumor was resected and an end-to-end anastomosis of the trachea was performed with non-resorbable sutures. Frozen sections of the tumor margins were free of malignant cells for at least 5mm. A thyroid lobe was wrapped around the anastomosis to protect it. Then, an orotracheal tube was placed within the reconstructed trachea and the patient was admitted to the critical care unit for 48 hours. Neck flexion was maintained for 5 days. The anastomosis was checked before the extubation with the fiber laryngoscope.

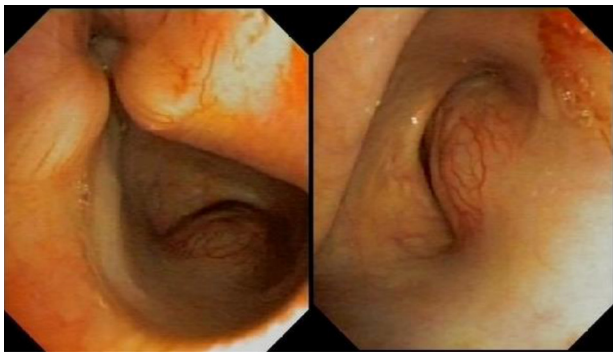

Figure 1: Bronchoscopy Showed the Mass Obliterating the Tracheal Lumen Below the Vocal Folds

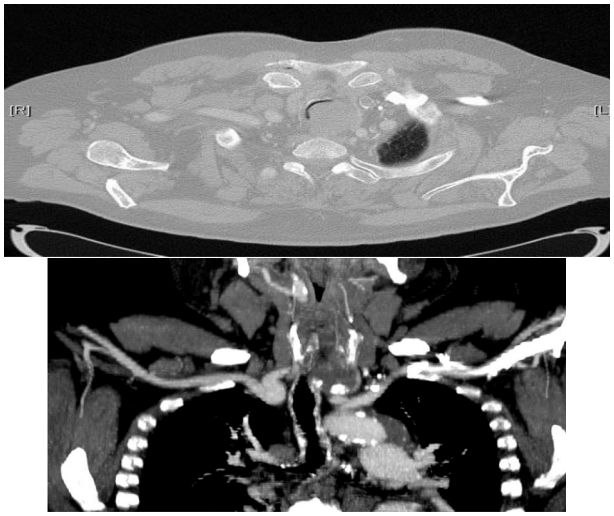

Figure 1: CT Scan Showed the Tumor in Axial and Coronal Planes

The specimen was 4.2cm of maximum diameter and it consisted of well-differentiated chondrosarcoma (grade I); showing a heterogeneous cellular pattern of regular eosinophilic cells with small nuclei with few multinucleated cells and mild atypia. Immunohistochemical study was positive for cyclin D1, RB, CDK4, and p53 staining; while it was negative for desmin, myogenin, S100, and p16 markers. The Ki-67 labeling index was below 25% which means low risk for metastasis and local relapse. Postoperatively, the patient was able to talk and breathe without the previous difficulty, and the anastomosis was assessed under direct vision with the fiber laryngoscopy; showing a correct mucosal healing and normal vocal cord function. The patient was discharged 2 weeks after the surgery due to a cardiac insufficiency presented within the postoperative period that required medical inpatient treatment. The follow-up of the patient was based on a control CT scan and a fiber laryngoscopy that revealed no recurrent disease. No adjuvant treatment was advised for this case and no complications related to the surgery such as tracheal stenosis or fistula were presented. The patient died without the recurrence of the underlying disease almost 7 years after the surgery, secondary to his cardiac and renal insufficiency.

DISCUSSION

Tracheal chondrosarcoma was first described in 1967 by Daniels et al., [5]. Since that time, only 18 cases have been described [3-19]; noting how

extremely rare this neoplasm is. Before that date, only 15 chondrosarcomas were described within the bronchial tree; while none of them were originated from the trachea. Chondrosarcoma is not such an infrequent tumor in other sites such as the pelvis, femur, or humerus; but it is scarcely found in the head and neck area (0.2% of the malignancies presented) [20]. It is slowly grown and has a slow onset with progressive dyspnea. The mass is usually asymptomatic until the mass occludes more than 75% of the tracheal lumen. This is why tracheal chondrosarcomas are often low-grade tumors with a diagnostic delay of one year and a half since the beginning of the symptoms [3]. Other common symptoms related to this tumor are non-productive cough and wheeze that may lead to a misdiagnosis of asthma not responsive to bronchodilators. Less frequently, it may debut with hemoptysis that accelerates the diagnostic process. This tumor predominantly affects males with only 1 female patient out of the 19 described cases [19]. The age of presentation is around the 6th decade of life, but there are 2 cases described at a much younger age; appearing in patients with Maffucci Syndrome [6, 8]. This syndrome is characterized by diffuse enchondromatosis and cutaneous haemangiomas distributed throughout the body, even though head and neck involvement is not so common [21].

These tumors are not often presented with metastasis. The only reported case presenting distant metastasis was the malignant transformation of an incomplete resected tracheal chondroma into high-grade chondrosarcoma which underlines the importance of complete resection of all benign tracheal masses [14]. In the majority of the cases (60%), tracheal chondrosarcomas affect the lower third of the trachea, whereas the superior and middle third account for 27% and 20% of the cases, respectively [3]. Computed tomography (CT) is the best imaging method to determine the location, tumor size, the extent of tracheal involvement, degree of luminal narrowing, and the evidence of extratracheal extension. Therefore it should be always performed before the elective surgery. Bronchoscopy is the gold standard modality to obtain a tissue sample that leads to a definitive diagnosis. It will not only lead to the histopathological diagnosis, but it will also allow performing endoscopic resection or laser debulking as a symptomatic treatment before surgery, or as a palliative treatment in cases for whom elective surgery is not advisable. Open surgical resection with negative margins and end-to-end anastomosis is the treatment of choice. This treatment is not accompanied by recurrence whereas all the patients that underwent an endoscopic resection were presented with recurrences [5, 11]. Maintenance of the airway is crucial during the interventions, and endoscopic debulking or mediastinal tracheostomy has been reported as a method to secure the airway, but the resection of tracheal chondrosarcoma with extracorporeal circulation has never been described [22]. Extracorporeal circulation allows maintaining the adequate amount of gas exchange and perfusion to sustain life; using extracorporeal membrane oxygenation of blood during the surgery stages where the trachea should be severed and the airway is at risk. It is a common technique used for cardiac surgery, but it has also been described as an alternative for endotracheal intubation, surgical crossfield intubation, and jet ventilation in tracheal resections [23]. Extracorporeal support contributes to ease the surgeon's maneuvers during the incision, resection, and reanastomosis of the trachea due to the absence of a ventilation device within the trachea and also avoids the risk of dislocation of the device during the surgery.

Radiotherapy has been described as an adjuvant therapy when endoscopic resection is performed [11] or when an open resection is contraindicated [7]. Since the majority of the tracheal chondrosarcomas are low-grade tumors, they are considered relatively radioresistant so that radiotherapy is not indicated in cases where negative surgical margins are achieved [24]. However, cases with residual disease after surgery and patients with unresectable tumors or not suitable for surgery may benefit from radiotherapy as palliative treatment. These malignancies are considered chemoresistant because of their slow rate of growth, variable matrix, and poor vascularization, and, therefore, chemotherapy is not advised except for high-grade chondrosarcomas. Our patient was the only case of tracheal chondrosarcoma that required extracorporeal circulation due to the tracheal lumen narrowing [25]. Maintenance of the airway was challenging and therefore the extracorporeal circulation was needed. It allowed performing the procedure safely and smoothly; allowing a complete

excision of the tumor that involved a long follow-up without recurrences.

Tracheal chondrosarcoma is a rare malignancy with only 19 cases described to date. It affects predominantly male patients in the sixth decade of life. Its main symptom is slow, progressive dyspnea that often misleads its diagnosis. CT scan allows to study its extension and to plan the surgery, but the definitive treatment is based on the histopathological study of a tissue sample obtained during a bronchoscopy. The treatment of choice is based on the open surgical resection of the tumor and end-to-end anastomosis, but maintenance of the airway results is challenging in many cases. We presented the only case of tracheal chondrosarcoma that required extracorporeal circulation during the resection procedure due to the tracheal lumen narrowing. Other modalities of treatment have resulted in disease recurrences and may only be advised as palliative treatments when the open elective surgery is not recommended. Radiotherapy may be used as adjuvant therapy, but only in cases with known residual disease or as a palliative treatment.

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None declared.

CONFLICTS OF INTEREST

The authors declared no conflict of interest.

ETHICS APPROVAL

All procedures performed in studies involving human participants were following the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from the participant included in the study.

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