

Tracheal condrosarcoma a rare tumor location. A case report

Condrosarcoma traqueal: una localización tumoral poco frecuente. Presentación de caso

Juan Pablo Manrique Salas[1]
Lina Marcela Quintero Díaz[2]
Francisco Enrique Barragán Araújo[3]
Mario Lemus Velázquez[4]
Anderson Rubén Lizcano Suárez[5]
Richard Eduardo Beleño Peñaloza[6]
https://doi.org/10.53903/01212095.307

Key words (MeSH)

Mediastinum
Chondrosarcoma
Tomography, X-Ray Computed
Trachea

Palabras clave (DeCS)

Mediastino
Condrosarcoma
Tomografía computarizada por Rayos X
Tráquea

Summary

Chondrosarcomas of the trachea are rare malignant tumors characterized by cartilage matrix synthesis. The annual incidence has been reported at 1 in 200,000 inhabitants/year. It has slow growth and is diagnosed late when it presents occlusions greater than 75% of the lumen of the trachea. Its diagnosis is based on images and endoscopic studies, and its management will depend on the stage, size, and extension to other structures. A case of tracheal chondrosarcoma is presented, along with the images and studies performed at the definitive diagnosis.

Resumen

Los condrosarcomas de tráquea son tumores malignos poco comunes que se caracterizan por la síntesis de matriz de cartílago. La incidencia anual se ha descrito en 1 de 200.000 habitantes/año. Tiene crecimiento lento y se diagnostica tardíamente, cuando tiene oclusiones superiores al 75% de la luz de la tráquea. Su diagnóstico se basa en imágenes y estudios endoscópicos; su manejo dependerá del estadio, tamaño y extensión a otras estructuras. Se reseña un caso de condrosarcoma traqueal, con las imágenes y estudios que llevaron al diagnóstico definitivo.

Introduction

Primary chondrosarcoma of the trachea is an extremely rare tumor, with few cases described in the literature; it originates from cartilaginous tissue and its etiology is still under study. Knowledge about this disease remains limited due to its uncommon nature (1).

These tumors occur most frequently in elderly men, are usually low grade and slow growing, with a low rate of recurrence and metastasis (2).

The most common symptoms are cough, dyspnea, and, less frequently, hemoptysis. Due to the slow growth of these tumors, patients often remain asymptomatic until the reduction in airway lumen is severe. Radiological diagnosis is usually performed with computed tomography (CT), which allows the extent and, frequently, calcifications to be documented. Fibrobronchoscopy is the gold standard diagnostic technique, useful for obtaining histological samples, for differential diagnosis, for airway clearance, and in evaluating tumor diameter and extent to define the therapeutic strategy (3).

Among the reported therapeutic alternatives are open resection with a lower recurrence rate, radiotherapy, stent placement, or the use of immunomodulators, depending on the stage of the disease and the possibility of resectability (2, 4).

Case description

A 65-year-old woman with no history of exposure to toxic substances or smoking; symptoms began five months ago with intermittent dry cough, followed by dysphagia for solids, which progressed to liquids and finally dyspnea. Physical examination revealed no visible abnormalities in the neck, with a symmetrical central trachea, no palpable adenomegaly or jugular engorgement, and no abnormal breath sounds.

An X-ray was performed, revealing an ovoid radiopaque image with defined edges, related to a mass in the upper right mediastinum (Figure 1). An esophagogram was also performed, showing threadlike passage of the contrast medium 20 to 26 cm from the dental arch (Figure 2). The third examination, a fiberoptic laryngoscopy, revealed erosive bulbar duodenitis and vocal cord paralysis. The fourth examination was a chest CT scan, which revealed a heterogeneous mass with lobulated, well-defined edges in the visceral (middle) mediastinum, surrounding and displacing the trachea anteriorly and to the left and the esophagus posteriorly, with images of calcific density inside it. After the ad-

[1]Resident physician specializing in Diagnostic and Therapeutic Imaging. Universidad Nacional Autónoma de México. Department of Diagnostic and Therapeutic Imaging, Hospital Centro Médico Nacional 20 de Noviembre. Ciudad de México, México. https://orcid.org/00000002-2245-9897

[2]Emergency medicine specialist. Universidad del Rosario, Universidad Industrial de Santander. Emergency Department. Clínica Chicamocha S.A. Bucaramanga, Colombia. https://orcid.org/0000-0002-8085-0464

[3]Resident physician specializing in Diagnostic and Therapeutic Imaging. Universidad Nacional Autónoma de México. Department of Diagnostic and Therapeutic Imaging, Hospital Centro Médico Nacional 20 de Noviembre. Ciudad de México, México. https://orcid.org/0009-0001-6894-891X

[4]Interventional Radiologist, Hospital General de México, Universidad la Salle. Department of Diagnostic and Therapeutic Imaging, Hospital Centro Médico Nacional 20 de Noviembre. Ciudad de México, México. https://orcid.org/00090006-1789-5332

[5]Orthopedic oncologist, Universidad Autónoma de México. Department of Oncology and Orthopedics, International Hospital of Colombia. Piedecuesta, Colombia. https://orcid.org/0000-00018245-0088

[6]Physician and surgeon. Universidad Industrial de Santander. Hospital Claudio Vicuña. Chile. https://orcid. org/0000-0002-0051-8251

ministration of intravenous contrast medium, avid and heterogeneous "flocculated" peripheral enhancement was observed inside (Figure 3). The fifth examination, upper gastrointestinal endoscopy, showed extrinsic obstruction in the upper esophagus with a 50% reduction in caliber (Figure 4a). Fibrobronchoscopy, the sixth examination, documented a solid tumor at the level of the 9th tracheal ring, on the lateral and posterior sides, with an 80% reduction in lumen and a 3 cm extension toward the carina (Figure 4b). A biopsy was obtained from two fragments of irregular soft tissue with a dark gray surface measuring 3 × 2 × 1 cm and 2 × 1 × 1 cm. The histopathological report showed a moderately hypercellular cartilaginous tumor with elongated nucleated cells and giant nuclei, sometimes binucleated, leading to a diagnosis consistent with chondrosarcoma.

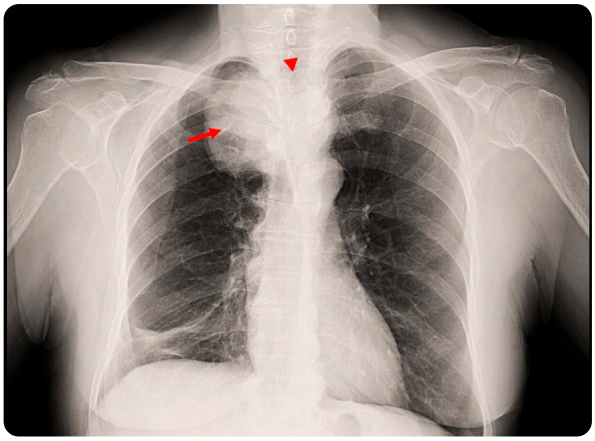


Figure 1. Chest X-ray. Heterogeneous lobulated mass (arrow) obliterating the middle segment of the trachea (arrowhead), lateral and posterior, with an 80% reduction in lumen and a 3 cm extension toward the carina (Figure 4b). A biopsy was obtained from two fragments of irregular soft tissue with a dark gray surface measuring 3 × 2 × 1 cm and 2 × 1 × 1 cm. The histopathological report showed a moderately hypercellular cartilaginous tumor with elongated nucleated cells and giant nuclei, sometimes binucleated, leading to a diagnosis consistent with chondrosarcoma.

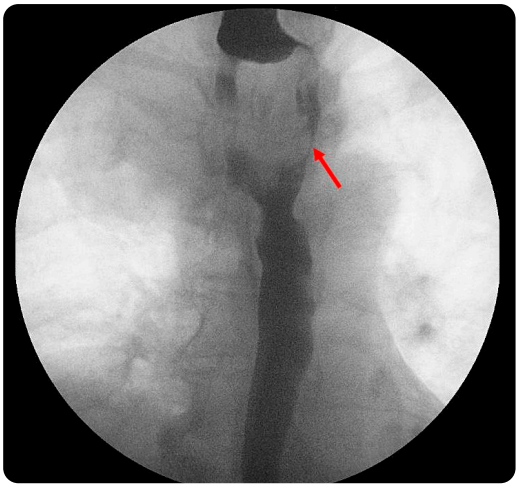


Figure 2. Anteroposterior view of esophagogram study with negative filling defect at the level of the upper esophagus, with threadlike passage of contrast medium in the lateral walls (arrow), 20-26 cm from the dental arch, suggestive of extrinsic esophageal compression.

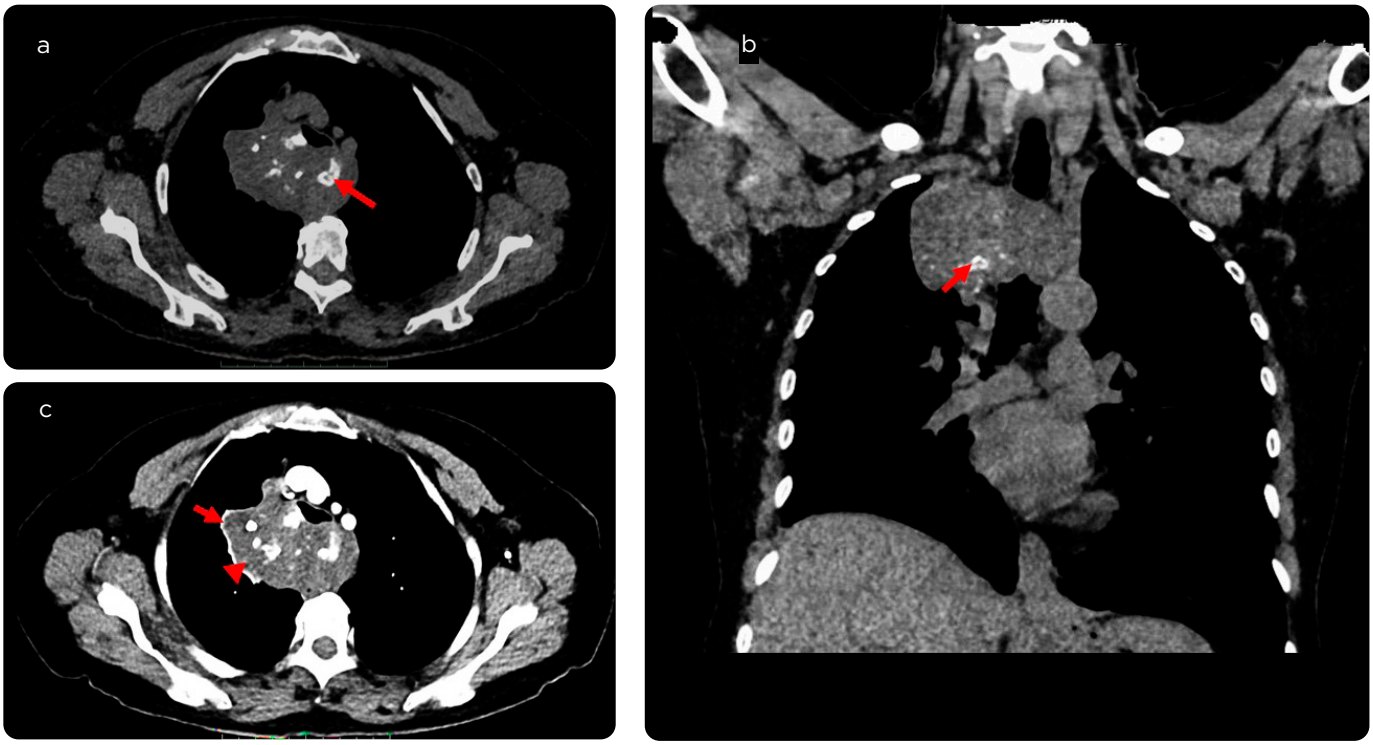


Figure 3. Chest CT scan. a) Axial and b) coronal, in simple phase: heterogeneous lobulated mass with defined edges, in the visceral mediastinum (middle), surrounding and displacing the anterior and left trachea and the esophagus toward the posterior mediastinum, images of calcium density inside (arrows). c) After intravenous contrast administration: avid peripheral enhancement (arrow) and heterogeneous "flocculated" enhancement inside (arrowhead).

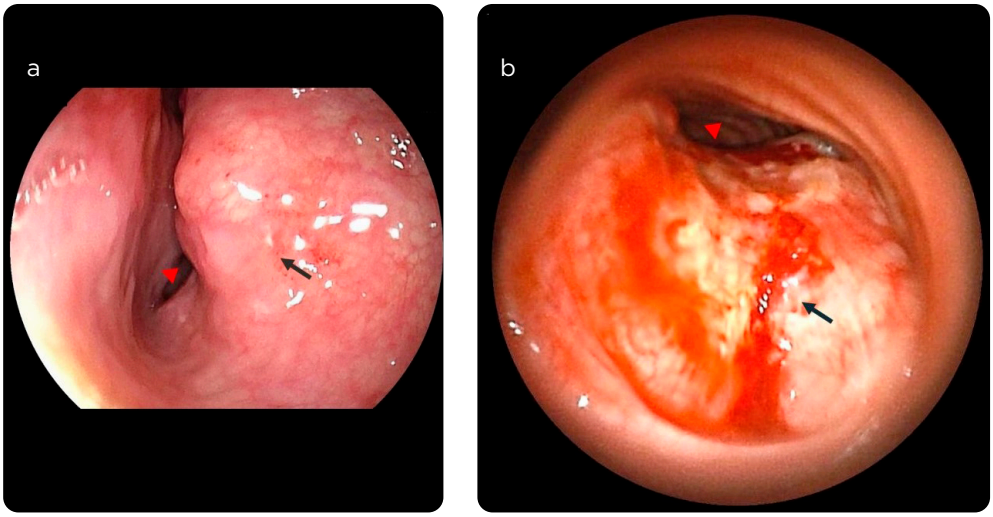


Figure 4. a) Upper gastrointestinal endoscopy: evidence of extrinsic obstruction at the level of the upper esophagus (arrow) that reduces its caliber by 50% (arrowhead). b) Bronchoscopy study: heterogeneous solid tumor with hemorrhagic areas (arrow), reducing the lumen by 80% (arrowhead).

Discussion

Chondrosarcomas are rare malignant tumors with an incidence of 1 in 200,000 inhabitants/year; they produce cartilaginous matrix and arise from the hyaline cartilage that covers the epiphyses of the bones. Lesions that arise de novo are called primary, and secondary lesions arise from chronic inflammation (2, 5).

Primary chondrosarcoma is the third most common malignant bone tumor and accounts for 20-27% of all primary malignant bone neoplasms. They frequently originate in the pelvis, femur, humerus, and ribs, followed by the head and neck in 10-12%, with the most common sites of origin being the larynx and maxillonasal region (5, 6).

Chondrosarcomas comprise only 0.1% of all head and neck tumors; they are usually low grade, slow growing, and lymph node (5%) and distant (7-18%) metastases have rarely been reported. Therefore, their impact is due to invasion and high recurrence rates (7).

Most cases arise from the bone tissue of the face and paranasal sinus cavities and, less frequently, from the cartilaginous tissues of the larynx or other soft tissues. Although the genesis of the tumor is not fully understood, it has been proposed that head and neck chondrosarcomas arise from the malignant degeneration of cartilage remnants, mesenchymal pluripotent cells, ossified cartilage, and isolated endochondromas; it has also been attributed to abnormal ossification of cartilage with chondroplasia due to chronic inflammation (8, 9).

Tracheal chondrosarcoma arises from the cartilage of the tracheal wall, predominates in the male population by up to 95%, and is most frequently located in the distal trachea (2). It slowly involves the tracheal lumen and surrounding tissues, causing obstructive respiratory syndrome; it is usually asymptomatic until the tumor occludes more than 75% of the tracheal lumen (10).

CT is the imaging modality of choice, as it allows determination of: size, location, presence or absence of calcification, percentage of luminal obstruction, and extratracheal extension. Radiological images show a soft tissue mass with scattered areas of calcification immersed in the chondroid matrix. Calcification is present in 71.4% of cases and extratracheal extension in 78.6%. It may or may not present central necrosis, and during CT with contrast medium, enhancement of the mesenchymal edge is observed, which has high diagnostic value; the type of enhancement is described as flocculent or streaked inside. On T1-weighted magnetic resonance imaging (MRI) images, it behaves with heterogeneous intermediate signal intensity, and after contrast administration, with septal and peripheral (edge) enhancement; on T2 sequences, scattered areas of high signal intensity are observed. These lesions contain abundant blood flow, giving them a "pepper surface sign" appearance (5).

Fibrobronchoscopy is the gold standard for diagnosis, as it provides therapeutic support through resection and/or reduction of tumor volume with laser (3). The use of endoprostheses in the airway may also be indicated for the treatment of tracheal chondrosarcomas when laser-assisted tumor reduction does not restore adequate airway patency or when airway collapse is observed (10).

Definitive open surgical treatment has a low recurrence rate, unlike that described in endoscopic resection; Adjuvant radiotherapy has been used in local recurrence after surgical excision in high-grade lesions to treat inoperable tumors for palliative purposes, in patients with incomplete resection or positive margins, in patients who are poor surgical candidates, and in patients who refuse margin clearance surgery (3).

Pembrolizumab has been approved for use in cases of unresectable lesions; however, a clear benefit for adjuvant therapy after resection and radiotherapy has not been established (3).

Tracheal chondrosarcoma is a rare, slow-growing tumor with an advanced clinical course at the time of diagnosis, because its symptoms are related to severe airway obstruction, which makes early diagnosis difficult and poses a diagnostic and therapeutic challenge for the physician; therefore, it is necessary to recognize the imaging characteristics to allow for early diagnosis.

References

1. Heuermann M, Bekker S, Czeczok T, Gregory S, Sharma A. Tracheal chondrosarcoma: A case report, systematic review, and pooled analysis. Cancer Rep (Hoboken). 2021:e1537. https://doi.org/10.1002/cnr2.1537. Epub ahead of print.
2. Kutzner EA, Park JS, Zaheer S, Inman JC. Tracheal chondrosarcoma: Systematic review of tumor characteristics, diagnosis, and treatment outcomes with case report. Case Rep Oncol Med. 2017;2017:4524910. https://doi.org/10.1155/2017/4524910
3. Thompson LDR, Gannon FH. Condrosarcoma de la laringe: un estudio clinicopatológico de 111 casos con una revisión de la literatura. Am J Surg Pathol. 2002;26(7):836-51. https://doi.org/10.1097/00000478-200207000-00002
4. Carretta A, Ciriaco P, Bandiera A, Muriana P, Arrigoni G, Negri G. Therapeutic strategy for tracheal chondrosarcoma: report of two cases. Monaldi Arch Chest Dis. 2020;90(1). https://doi.org/10.4081/monaldi.2020.1223
5. Murphey MD, Walker EA, Wilson AJ, Kransdorf MJ, Temple HT, Gannon FH. Imaging of primary chondrosarcoma: Radiologic-pathologic correlation. RadioGraphics. 2003;23:1245-78. https://doi.org/10.1148/rg.235035134
6. Hong P, Taylor SM, Trites JR, Bullock M, Nasser JG, Hart RD. Condrosarcoma de cabeza y cuello: informe de 11 casos y revisión de la literatura. J Otolaryngol Head Neck Surg. 2009;38(2):279-85. https://doi.org/10.2310/7070.2009.080139
7. Ruark DS, Schlehaider UK, Shah JP Condrosarcomas de cabeza y cuello. Rev Mund Cirug. 1992;16(5):1010-5. https://doi.org/10.1007/bf02067021
8. Wagnetz U, Patsios D, Darling G, Las Heras F, Hwang D. Condrosarcoma traqueal: una complicación poco frecuente en el síndrome de Maffucci. Br J Radiol. 2009;82(981):e178-e181. https://doi.org/10.1259/bjr/17386896
9. Bhat AM, Gerndt S, Sandhu A, Helfand AM, Jiwani R, Low GMI. Tracheal chondrosarcoma. A novel presentation of Lynch syndrome. JAMA Otolaryngol Head Neck Surg. 2024;150(2):183-4. https://doi.org/10.1001/jamaoto.2023.3908
10. Rotolo N, Cattoni M, Nardecchia E, De Maio S, Franzi F, Pettenon F, et al. Surgical management of tracheal condrosarcoma. Acta Chirurg Belgica. 2020;25. https://doi.org/10.1080/00015458.2020.1863039

Correspondence

Juan P. Manrique-Salas
Avenida Félix Cuevas 540, Colonia del Valle Sur, Benito Juárez
Código postal: 03229
Ciudad de México, CDMX, México
jpmansa@gmail.com

Received for evaluation: March 11, 2024
Accepted for publication: April 19, 2024