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Short Communication

Chondrosarcoma of the upper third of the trachea: Case report and review of the pathology features

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ABSTRACT

Malignant tumors of the trachea are rare. The most frequent histologic type is squamous cell carcinoma. Cartilaginous tumors are rare in the upper airways. There are only four reported cases of chondrosarcomas of the upper third of the trachea. We present a 76-year-old man with an upper third tracheal chondrosarcoma and a review of the literature, focusing on the clinicopathological features.

KEYWORDS: chondrosarcomas, chondromas, trachea, cartilaginous tumors

ABBREVIATIONS

CT scan : computed tomography scan
ED : emergency department

INTRODUCTION

Cartilaginous tumors that compromise the airways are rare. From these, chondrosarcomas hardly ever occur having only eighteen cases reported in the literature between 1954 and 2009.

And only four cases involving the upper third tracheal portion have been described.

We present a 76-year-old man with an upper third tracheal chondrosarcoma and a literature review, focusing on the clinicopathological features.

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CASE REPORT

A 76-year-old, obese man who had several emergency department (ED) visits due to syncopal episodes in 2008. At one of his last visits, a chest radiography was performed observing a narrow tracheal lumen (Fig. 1A). Later on, a CT scan was done identifying a 23 mm - upper third tracheal portion mass with multiple calcifications (Fig. 1B). A required bronchoscopy revealed a tumour covered by normal mucous membrane without ulceration that occluded 50% of the tracheal lumen. Three years later, the patient presented again at the ED with progressive dyspnea without cough or expectoration. A new CT-scan was performed, showing a 45 mm calcified mass. There was no evidence of lymph nodes. A new bronchoscopy revealed an enlargement of the prior tracheal mass occluding more than 90% of its lumen, covered by a hypervascular mucosal membrane (Fig. 1C).

Due to his age, obesity and medical history of thoracic aortic aneurysm, a tumor debulking was performed.

The pathological approach revealed a well differentiated, hypercellular cartilaginous neoplasm with proliferating pattern that consisted of clusters of cells with nuclear hypercromasia, showing frequent binucleated cells and calcifications. Mitotic figures were not seen and proliferation index performed by Ki67 was 0.5-1% (Fig. 1D).

DISCUSSION

Tracheal malignant tumors are rare. The most frequent histologic pattern is squamous cell

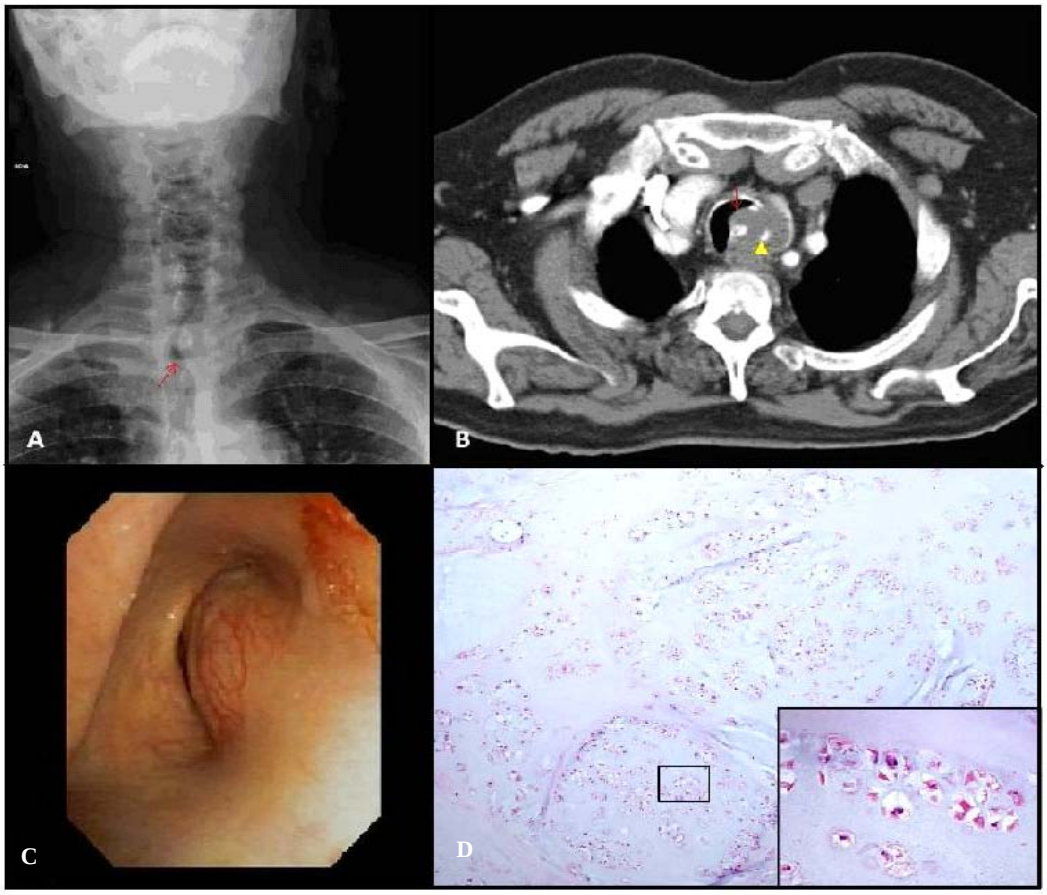

Fig. 1. A) The chest radiograph shows a narrow cervical tracheal lumen (arrow). **B**) CT scan with intravenous contrast shows a 45 mm mass with calcifications on its greatest diameter. The tumor depends on the left posterolateral tracheal wall and significantly narrows the airway. **C**) Endoscopic image showing the most cranial part of the tumor obstructing almost all of the tracheal lumen. **D**) Moderately hypercellular cartilaginous neoplasm with proliferating pattern. Inset: clusters of cells with round and polygonal cytoplasm, nuclear hypercromasia, showing frequent binuclear cells.

carcinoma (60-90%) followed by adenoid cystic carcinoma. The remaining tracheal malignancies are very infrequent, which include chondrosarcoma, leiomyosarcoma, fibrosarcoma, spindle cell sarcoma, synovial cell sarcoma, and primary melanoma [1].

Chondrosarcomas can occur anywhere but are more common among long bones and pelvis [2]. Cartilaginous tumors of the trachea very seldom occur. Chondrosarcomas are less frequent than chondromas. Therefore only 18 cases of tracheal chondrosarcomas have been described on a literature review [1, 3-19]. We provide the nineteenth case, whose clinical and pathological features are summarized in Table 1.

Chondrosarcomas frequently affect adults between the sixth and ninth decade of life with an age range among 32 and 87 years. Of the reported cases there was male predominance, describing only two female cases [6, 18].

Patients presented upper-airway obstruction symptoms when more than 75% of the tracheal lumen was occluded: dyspnea, wheezing and stridor. These symptoms were presented 1 to 36 months before diagnosis. In some cases there were mucosal irritation and ulceration with cough and hemoptysis [17, 19]. Dyspnea may progress and get worse rapidly, as in our patient. The combination of cough, wheezing, and dyspnea without hemoptysis often leads to misdiagnosis

Table 1. Review of all published cases in literature including our case.

Authors	Age/ sex	Symptoms/ duration (months)	Location	Size	Treatment		Pathology	Recurrence	Prognosis (years)
					Endoscopy	Surgery			
Moersch[3] 1954	NR	NR	NR	NR	NR	Resection	NR	NR	NR
Jackson[4] 1959	32/M	C/72	L	NR	Resection	ND	NR	No	NED; 6.0
Daniels[5] 1967	73/M	C, W, D/7	L	2.5	ND	ND	NR	Local	AWD; 2.5
Fallahnejad[6] 1973	48/F	C, W, D/16	U	4.0	ND	7 rings	Ex: Extratracheal Ca: No	No	NED; 5
Weber[7] 1978	71/M	H, C, D/4	M	3	Resection	No	Ex: Extratracheal Ca: Present	No	NED; 5.0
Slasky[8] 1985	58/M	D/24	L	2	ND	4.0 cm	Ex: Extratracheal Ca: Present	No	NED; 2.5
Arevalo[9] 1986	74/M	P, A/NR	U	2	ND	2.5 cm	Ex: Extratracheal Ca: Present	No	NED; 1.0
Matsuo[10] 1988	72/M	H, D/7	M	5	Debulking	4.5 cm	Ex: Extratracheal Ca: No	No	NED; 0.5
Salminem[11] 1990	54/M	W, D/1	L	2	ND	Incomplete	Extension: NR Calcification: No	Loc + dist	DOD; 14.0
Mine[12] 1990	74/M	C, D/6	L	4.5	Debulking	6 rings	Ex: Extratracheal Ca: NR	No	NED; 2.8
Kaneda[13] 1993	64/M	D/36	L	2.3	ND	3 rings	Ex: Tracheal Ca: NR	No	NED; 2.8
Leach[14] 1994	78/M	D/36	L	6.5	ND	3 cm	Ex: Extratracheal Ca: Present	No	NR
Kiriyama[15] 1997	54/M	W, D/1	L	2	Debulking	6 rings	Ex: NR Ca: Present	No	NED; 3.5
Farrel[16] 1998	87/M	D/12	M	3	Resection	ND	Ex: Extratracheal Ca: NR	Local	AWD; 1.0

Table 1 continued..

Maish[1] 2003	78/M	D/3	L	NR	Debulking	Resection	Ex: Extratracheal Ca: No Well encapsulated	No	NED; 2.5
Umezu[17] 2008	34/M	H, W, D/8	U	2.5	ND	5 rings	Ex: Extratracheal Ca: Present	No	NED; 6.3
Hong[18] 2009	77/F	NR	NR	NR	NR	Resection	NR	NR	NR
Mendoça[19] 2010	72/M	O, D, W 12	U	NR	Debulking	ND	Ex: Extratracheal Ca: Present	No	NED; 7
López-Rubio 2011	76/M	D/10	U	4.5	Debulking	ND	Ex: Extratracheal Ca: Present	No	-

C: cough; W: Wheezing; D: dyspnea; H: haemoptysis, P: pneumonia; A: asymptomatic; O: odynophagia; L: lower; M: medium; U: upper; ND: not done; Ca: calcification; Ex: extension; DOD: death of disease; NED: no evidence of disease; AWD: alive with disease; NR: not reported.

with asthma or chronic obstructive pulmonary disease [1].

The initial evaluation commonly involves chest radiography that is frequently normal. A CT-scan is the gold standard imaging method. It lets us evaluate the tumor characteristics such as its localization, size, calcification, obstruction degree, extra-tracheal compromise, resectability, nodal involvement and distant metastases [1]. 8 of 19 patients showed multiple calcifications in the CT scan and 18 had extratracheal extension [15]. Bronchoscopy is the best method to obtain a biopsy for diagnosis and it is also useful for tracheal obstruction treatment.

Of the 19 cases, 9 were located in the lower third of the trachea, 3 in the middle third, and 5 in the upper third, including our case. The localization of the remaining case is unknown. The tumor size ranged from 2.0 to 6.5 cm in diameter. Tracheal chondrosarcomas can be staged in grades I, II and III as bony chondrosarcomas [20]; because there is a direct correlation between its histological grade and prognosis. Tumor grade is determined by cellular density, mitotic figures frequency, atypia, presence and necrosis extension. High-grade tumors may have a spindle cell appearance and an increasingly myxoid matrix. The majority of tracheal chondrosarcomas have low-grade differentiation in comparison with other body location ones, which are more aggressive [18].

The distinction between chondroma and low-grade chondrosarcoma is very difficult. The pathological features are similar, presenting cellular atypia, binucleated cells and calcifications [1, 17, 18, 19]. Therefore, Weber *et al*. proposed chondroma and chondrosarcoma as a single entity: “cartilaginous tumors”. However, chondromas are usually small lesions (<1-2 cm) with intraluminal extension, whereas chondrosarcomas present extraluminal growth. High-grade chondrosarcomas are more aggressive, with regional and distant metastasis. Those involving the trachea tend to have slow growth and do not metastasize. Skeletal chondrosarcomas have a more aggressive behavior.

Salminen *et al*. [11] presented a case of an incomplete resected chondroma that had several recurrences. Fourteen years later, multiple lung metastases were presented and the patient died. Due to this, recurrences and malignant transformation are the most feared complications, justifying a follow-up of 10 years minimum.

CONCLUSION

Tracheal chondrosarcoma is a very rare presentation, with slow growth rate and an indolent course (low-grade chondrosarcomas). Differential diagnosis between chondromas and chondrosarcomas is sometimes difficult. It is accepted that chondromas have no extratracheal extension, up against chondrosarcomas. Otherwise, the reported case of recurrent chondroma [11] without complete resection suggests that tracheal chondromas should be considered as chondro-sarcomas because of their potential for malignant transformation.

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