

## Chondrosarcoma of the thyroid cartilage

KEMAL UYGUR, M.D., MUSTAFA TÜZ, M.D., HARUN DOĞRU, M.D., ALIYE SARI\*

### Abstract

Sarcomas of the larynx are extremely rare neoplasms that account for approximately one per cent of all tumours of this organ. We present a case of laryngeal chondrosarcoma originating from thyroid cartilage, characterized by an unusual long clinical course over 15 years without laryngeal symptoms or duplication of metastases, treated at the Süleyman Demirel University Medical Faculty, Ear, Nose and Throat (ENT) department.

**Key words:** Thyroid Cartilage; Chondrosarcoma

### Introduction

Although chondrosarcoma is rarely encountered in the head and neck, the larynx is the most frequent site of origin of chondrosarcoma when it does occur. Further, chondrosarcoma is the most frequent sarcoma of the larynx.<sup>1</sup> Although all sarcomas of the larynx are extremely rare neoplasms, accounting for only about one per cent of all tumours of the organ.<sup>2</sup> Of these, less than 0.1 per cent are chondrosarcoma.<sup>3</sup> Two hundred and twenty cases of this disease entity have been reported in the world literature.

In 75 per cent of chondrosarcoma cases the site of involvement was the cricoid cartilage, while 20 per cent have involved the thyroid cartilage. The tumour presents either as a space-occupying lesion in the subglottic region or as a neck mass.

Chondrosarcomas of the larynx tend to be slow-growing and to behave in a less aggressive manner than tumours of the same histological type of extra-laryngeal origin.<sup>4</sup> Radiotherapy and chemotherapy are of little value and surgical removal is the widely accepted treatment.<sup>5</sup>

We present a case of laryngeal chondrosarcoma originating from the thyroid cartilages, characterized by an unusual long clinical course of over 15 years without laryngeal symptoms or the development of metastases, treated at the Süleyman Demirel University Medical Faculty, ENT department.

### Case report

A 77-year-old man presented in November 1999 for evaluation of a firm non-tender right-sided neck mass that had reportedly enlarged over a 15-year period. He had no history of dysphagia or hoarseness. He had pain in the neck for two months. Examination revealed a 7 × 6 cm fixed right anterior triangle mass with no evidence of adenopathy. On indirect laryngoscopy, the mucosa of the right piriform sinus appeared intact but there was a little medial displacement. No evidence of endolaryngeal involvement was observed.

Computerized tomography (CT) demonstrated a large expansile lesion arising from the right lamina of the thyroid cartilage. Areas of stippled calcification consistent with a



Fig. 1

Coronal contrast CT scan demonstrates stippled calcification within a large mass arising from the thyroid cartilage ala and extending into the extralaryngeal tissues.

chondroid matrix were noted. Tumour margins were well-defined. The mass rated as hyperdense compared with adjacent muscle (Figure 1). Linear contrast absorbance was seen on the periphery of the lesion. The airway was slightly displaced medially. No evidence of adenopathy was noted. The diagnostic impression subsequent to CT evaluation was probable chondroma or chondrosarcoma. The pathological findings from a fine needle aspirate also suggested chondroma or well-differentiated chondrosarcoma. CT of lung and bone scintigraphy revealed no evidence of metastasis.

The patient was taken to the operating theatre. A horizontal 10 cm right mid-cervical incision was done. Following delineation of the encapsulated mass, it was dissected from the surrounding tissue. It was seen that the

From the Departments of Otolaryngology – Head and Neck Surgery and Pathology\*, Süleyman Demirel University School of Medicine, Isparta, Turkey.

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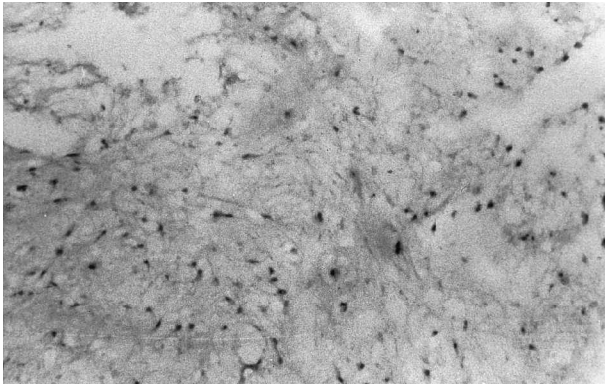


FIG. 2

Low-power view of a laryngeal chondrosarcoma showing lobulation (H&E;  $\times 40$ ).

mass originated from the right thyroid ala. Both superior and inferior cornua of the right thyroid ala were dissected free. The right thyroid ala was separated from the left by midline incision. The medial perichondrium was dissected. This dissection allowed for removal of the right thyroid ala with the tumour, which appeared to be intact. The strap muscles were re-approximated superiorly, and the wound was closed following placement of a suction tube. The peri-operative and post-operative periods were uneventful. The histopathological examination of the operation specimen revealed areas of increased cellularity and a greater degree of cytological abnormality. Neoplastic cells formed clusters of irregular size and distribution (Grade 2, chondrosarcoma with myxoid components) (Figures 2 and 3). The patient was followed up for a period of eight months. He had no hoarseness pre-operatively nor post-operatively.

### Discussion

Although chondrosarcoma is the most common mesenchymal tumour of the larynx, there have only been approximately 220 cases reported.<sup>6</sup> The majority of the tumours arise from the cricoid cartilage. The posterior lamina is the most frequent site within this cartilage.<sup>7</sup> Although the thyroid cartilage is the next most common site of origin, tumour arising from thyroid cartilage is reported to be infrequent. Fifty per cent of chondrosarcomas of the thyroid cartilage originate from the external surface of the thyroid lamina.<sup>7</sup> These tumours most commonly affect adults between 40 and 60 years of age,<sup>2,8</sup> with a male-to-female ratio of 4:1.<sup>8,9</sup>

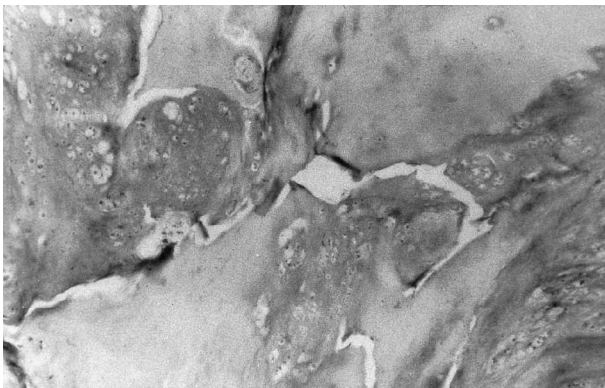


FIG. 3

Closer view of cords and strands of round, slightly elongated cells embedded in a myxoid stroma (H&E;  $\times 100$ ).

Symptoms related to cricoid involvement result from the slow intraluminal growth of the lesion in the subglottic area. At indirect laryngoscopy, typical cricoid lesions appear as a mass covered by healthy-looking mucosa, occupying the subglottic region. There may be impaired movement of one or both vocal folds. This produces dyspnoea, hoarseness and eventually dysphagia. If the tumour is located in the thyroid cartilage, it may present as a cervical mass. In our case no laryngeal symptom was detected, as the tumour arose from the external surface of the right lamina of the thyroid cartilage, and the patient was admitted to our clinic complaining of a mass on the right side of the neck.

Roentgenographically, on plain lateral and anteroposterior films, these lesions may show a smooth mucosal outline with calcification and either peripheral or central stippling and ossification.<sup>10</sup>

Computed tomography is a valuable radiological tool in the evaluation of laryngeal tumours. One advantage of CT over conventional radiological techniques is the superior resolution and visualization of the larynx in the transaxial plane. In the case of laryngeal chondrosarcomas, CT reveals areas of calcification in 80 per cent of cases.<sup>11</sup> These calcifications are characteristic for this tumour. However, the differential diagnosis based on clinical findings is so broad that histological examination is the only means of definite diagnosis.

Criteria for pathological diagnosis of chondrosarcoma include the presence of many cells with large, irregular and/or multiple nuclei, and giant cartilage cells with large single or multiple nuclei and nuclei containing clumped chromatin.<sup>12</sup>

Laryngeal chondrosarcomas are typically low grade malignancies and tend to be much less aggressive than chondrosarcomas that arise elsewhere. These tumours respond poorly to both chemotherapy and radiotherapy, and these methods are not regarded as treatment options. Surgical excision is the treatment of choice, and since local and intrinsic spread is the rule and metastatic spread is uncommon, conservative resection is considered the best approach in most circumstances. Conservative surgery was performed in our case as no locoregional metastases had been detected, though he had a 15-year history.

Long-term prognosis depends upon tumour grading, location and initial extent, and the adequacy of the first surgical procedure.<sup>5</sup> High-grade chondrosarcomas recur locally and tend to metastasize to lung or bone; low-grade tumours do not normally metastasize.<sup>13</sup> Our case was a Grade II chondrosarcoma (with myxoid components) and the case is being followed up for eight months for metastases and recurrence.

Although chondrosarcoma arising from the thyroid cartilage is extremely rare, it should be considered in the differential diagnosis of solid or cystic neck masses. Differential diagnosis should include other primary laryngeal lesions such as neurilemmoma, adenocarcinoma, amyloid tumour, granular cell myoblastoma, plasmacytoma, advanced squamous cell carcinoma of the larynx, secondary or metastatic laryngeal tumours and chondrometaplasia.<sup>5</sup>

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Address for correspondence:  
Kemal Uygur, M.D.,  
PK: 132 Isparta,  
Turkey.

Fax: (246) 2371758  
E-mail: mutuz@hotmail.com

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