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Chondrosarcoma of the larynx

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Abstract

A case of a low grade chondrosarcoma of the cricoid cartilage which had been diagnosed initially as a chondroma is presented. The tumour recurred twice after limited surgical resections. Total laryngectomy was inevitable due to near total involvement of the cricoid cartilage and subsequent histological examination revealed a low grade chondrosarcoma. We have discussed in brief, the diagnosis and treatment of chondrosarcomas of the larynx and support the view of conservative surgical management for low grade tumours as they are slow growing and metastases are infrequent. A total laryngectomy may be reserved for salvage or primarily when more than half of the cricoid cartilage needs to be resected.

Histological grading reveals the biological behaviour of the tumour and CT scans help in planning the surgery. A regular follow-up is necessary for early detection of recurrences and metastases.

Key words: Laryngeal neoplasms; Chondrosarcoma; Laryngectomy

Introduction

Chondrosarcomas of the larynx are uncommon with approximately 204 cases having been reported in the literature (Ferlito *et al.*, 1984; Nicolai *et al.*, 1990). These tumours are usually slow growing with low grade malignancy (Finn *et al.*, 1984; Burkey *et al.*, 1990). Regional and distant metastases are rare (Nakayama *et al.*, 1993) and treatment varies from a limited resection to a total laryngectomy depending on the site, extension and histological grade of the tumour. We present a case report and discuss in brief the diagnosis and management of chondrosarcoma of the larynx.

Case report

A 53-year-old man was first seen in June 1980, at a local clinic for evaluation of hoarseness. Examination revealed a subglottic mass with impaired mobility of the right vocal fold. A mass contiguous with the posterior lamina of the cricoid cartilage was noted in the soft tissue X-ray studies. The tumour was excised through an external approach and the biopsy was reported as benign chondroma. Two years later, the patient was again re-evaluated elsewhere for persistent hoarseness and progressive dyspnoea. The patient underwent tracheostomy. Direct laryngoscopy and CT scans revealed a subglottic mass involving the right posterolateral portion of the cricoid cartilage. The recurrent tumour was resected through an external approach and the histological examination revealed a benign chondroma.

The patient was referred to our department in September 1989, for evaluation of dysphagia, aphonia and a neck swelling. The tracheostomy tube was in place and the fibroscopic examination of the larynx revealed a smooth bulging mass encroaching the laryngeal lumen anteriorly and the hypopharynx posteriorly. The CT scans showed a hypodense mass occupying more than half of the cricoid cartilage with mottled calcification (Figure 1). The roentgenograms of the chest were normal. A total laryngectomy and a right hemithyroidectomy were performed.

The gross examination of the surgical specimen revealed a tumour (6 cm diameter) replacing the posterior lamina of the cricoid cartilage. Microscopic examination showed a well differentiated cartilaginous tissue with a typical cell nuclei. The cytological features included double nucleated chondrocytes, anisocytosis, anisokaryosis and absence of mitoses (Figure 2). The tumour did not penetrate through the perichondrium, and the paralaryngeal soft tissues were not invaded. The final histological diagnosis of this recurrent tumour was a low grade chondrosarcoma.

The post-operative course was uneventful. The patient developed a good oesophageal voice and he is currently alive with no evidence of local or distant disease five years after the salvage laryngectomy.

Discussion

Chondrosarcomas of larynx comprise less than one per cent of all malignant tumours of the larynx (Batsakis and Raymond, 1988). The cricoid cartilage is the most commonly affected site with a special predilection for the posterior lamina (Neis *et al.*, 1989). The other sites involved in descending order of frequency are thyroid cartilage, arytenoid, vocal fold and epiglottis (Cocke, 1962; Gasior and Remine, 1967; Barsocchini and McCoy, 1968). These tumours occur more frequently during the sixth or seventh decade of life with a striking male preponderance.

The symptomatology depends on the site of origin and extent of the tumour. Dyspnoea and hoarseness of voice are frequent with cricoid lesions and dysphagia occurs when the tumour grows posteriorly towards the hypopharynx. Vocal fold paralysis is almost exclusively an early sign of cricoid lesion and may be related to involvement of the recurrent nerve or fixation of the cricoarytenoid joint (Ferlito, 1993).

The CT scans were helpful in determining the site and extent of the lesion. Mottled calcification was reported in 80 per cent of soft tissue X-ray studies (Zizmor *et al.*, 1975) and, if present, is pathognomic of this type of tumour.

Endoscopic examination may reveal a smooth, mucosa covered mass in the posterior or posterolateral subglottic region. Sometimes biopsy may be difficult or impossible due to hardness

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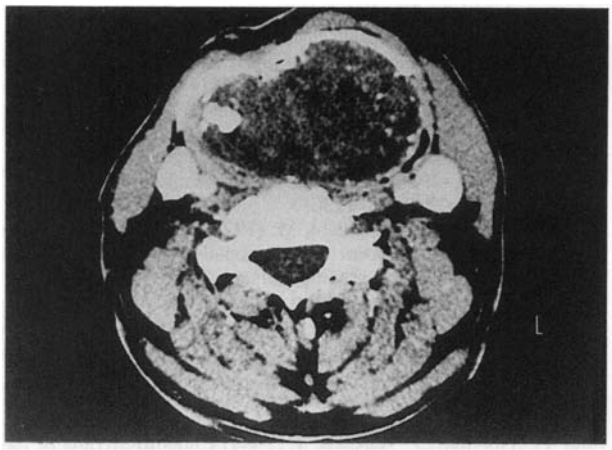

FIG. 1

CT scan of the larynx showing a hypodense lesion of the cricoid cartilage with areas of mottled calcification.

of the lesion. The fragmented biopsy may not display proof of malignancy.

The histological pattern of chondrosarcoma varies from a well differentiated growth that can be similar to benign chondroma to a high grade malignancy that exhibits aggressive local behaviour and potential for metastases (Finn *et al.*, 1984). By definition, a benign chondroma histologically duplicates the normal cartilage, be it elastic or hyaline in type, and may exhibit an increased cellularity but the individual cells retain a uniform structure similar to that in normal cartilage (Neis *et al.*, 1989). Lichenstein and Jaffe (1943) laid down the histological criteria for chondrosarcoma and reported that the cartilage tumour should no longer be regarded as benign if, when viable noncalcifying areas are examined it shows, even in scattered fields: (1) many cells with plump nuclei, (2) more than one cell with two such nuclei, (3) giant cartilage cells with large single or multiple nuclei or with clumps of chromatin. Evans *et al.* (1977) further divided chondrosarcomas into grades I to III (low, medium and high grades) based on the number of mitoses, and nuclear, cellular and architectural atypia. The histological characteristics of grade I chondrosarcoma include, (a) abundant chondroid to myxoid martix, (b) predominence of cells with small and densely staining nuclei, (c) binucleated or occasionally multinucleated cells, (d) frequent calcification or sometimes bone formation, (e) dense cellularity with nuclear irregularity and predominence of nucleoli, (f) mitosis is rare to absent (Burkey *et al.*, 1990; Ferlito, 1993).

Medium and high grade tumours show obvious anaplasia. But, the histological differences between a chondroma and a grade I chondrosarcoma are subtle, resulting in frequent misdiagnosis of the malignancy in the absence of other clinicoradiological and pathological data. Batsakis and Raymond (1988) emphasized that the presence of clustering of cells (cluster disarray) in the low grade tumours is the significant histological difference from chondroma. There is an overlap of cytological similarities between these two lesions, and there are often difficulties in histological differentiation based on Jaffe's double nucleated chondrocyte, hypercellularity and general plumpness. About 25 per cent of chondrosarcomas are so well differentiated that anaplasia may be absent and these low grade tumours are frequently amitotic. The double nucleated chondrocytes can be also seen in benign cartilaginous tumours. Mirra (1989) noted that the probability of chondrosarcoma is 90 per cent or greater if the number of double nucleated cells exceeds 26 per 20 high power fields. He measured also the cellularity of these two lesions and found that the areas of maximal cellularity correlated to malignancy. The average maximum cellularity was 200 cells × 400 field for grades II and III chondrosarcomas, 100

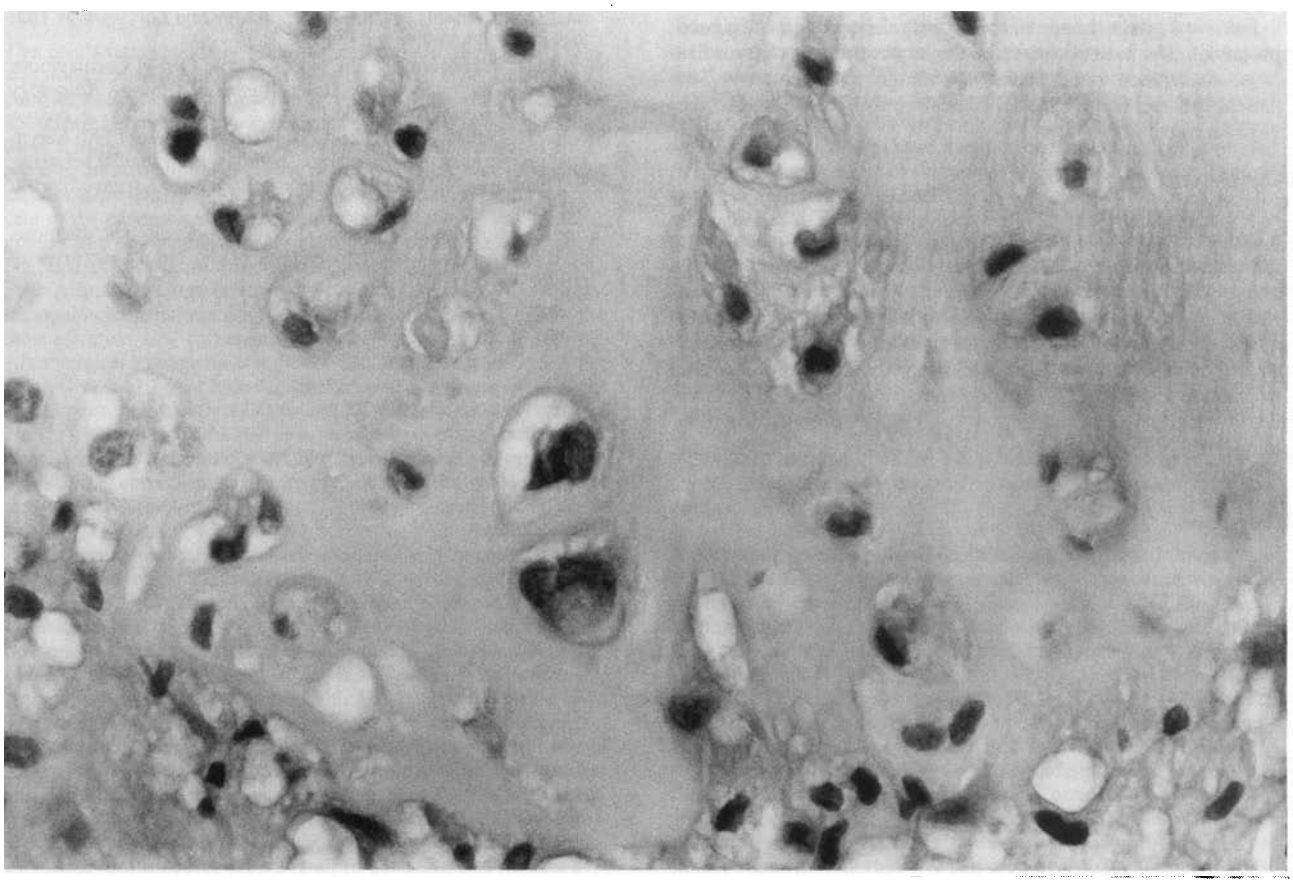

FIG. 2

Well differentiated low grade chondrosarcoma showing nuclear atypia and binucleate cells. (H & E; × 400).

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cells × 400 field for grade I chondrosarcomas, whereas the cellularity of chondromas was one half that of low grade chondrosarcomas.

Regional and distant metatases are rare. To date, approximately 20 cases of regional and distant metastases from laryngeal chondosarcomas have been reported in the literature (Nakayama *et al.*, 1993) with an estimated incidence of 10 per cent. The Lung was the most common site followed by cervical lymph node, kidney, bone and subcutaneous nodules.

Surgery remains the mainstay of treatment for chondrosarcomas. Harwood *et al.* (1980) stated that chondrosarcomas are potentially radiocurable and suggested post-operative radiation for unresectable disease or inadequate surgical margins. The role of chemotherapy as adjuvant in the initial treatment is controversial. However, chemotherapy may offer a palliation for chondrosarcoma with aggressive local spread or metastases (Finn *et al.*, 1984).

The distinction between chondroma and low grade chondrosarcoma is often difficult and the diagnosis of malignancy is often established at recurrence. However, the initial distinction between the two is helpful only in predicting the biological behaviour of the tumour. Lavertu and Tucker (1984) found that the eventual outcome of the cases initially labelled as chondroma was not significantly different from those identified as chondrosarcoma at the outset. Because of the slow growth and low metastatic potential of low grade chondrosarcomas, conservative surgery has been suggested initially or on recurrence, up to the point where an adequate airway cannot be reconstructed (Lavertu and Tucker, 1984). A total laryngectomy should be considered initially when the tumour involves more than half of the cricoid cartilage (Barsocchini and McCoy, 1968) or in the cases of histologically high graded or dedifferentiated tumours (Jones, 1973; Nakayama *et al.*, 1993). Since nodal metastases to the neck are rare, elective neck dissections are not indicated (Hicks *et al.*, 1982).

In our case, the tumour was underdiagnosed initially, and was in fact a low grade chondrosarcoma with slow growth and a good prognosis. The patient conserved the larynx for nine years until a large recurrence necessitated a salvage laryngectomy. The patient is currently alive and disease-free five years after the salvage laryngectomy.

Conclusions

We have reported a case of low grade chondrosarcoma of the cricoid cartilage. The growth of low grade tumours is slow and metastases are infrequent. The histological distinction from a benign chondroma is always difficult. We believe that conservative surgery should be employed initially or later as salvage, if an adequate airway can be reconstructed. Total laryngectomy should be reserved for large primary or recurrent tumours and high grade lesions. Adjuvant radiotherapy should be considered for involved surgical margins or high grade tumours. Regular, long-term, follow-up is necessary to detect recurrences and metastases.

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