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CHONDROSARCOMA OF THE HEAD AND NECK

Muzaffer KANLIKAMA *

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SUMMARY

Chondrosarcoma is slow growing but locally aggressive malignancy with a propensity for progressive spread and multiple recurrences that eventually leads to death if inadequately treated, and uncommonly seen in the head and neck.

In this article, a review of literature regarding on the chondrosarcoma of the head and neck was presented and treatment modalities and results o this alternatives were discussed.

ÖZET

BAŞ-BOYUN KONDROSARKOMLARI

Baş-boyun bölgesinde sık görülmeyen kondrosarkomlar; yavaş büyüyen fakat lokal olarak agresif ve lokal nüks eğilimi yüksek olan ve uygun tedavi edilmedikle-rinde sonuçta ölüme yolaçabilen tümörlerdendir.

Bu makalede, baş-boyun kondrosarkomları gözden geçirilmiş ve tedavi yöntemleri ile bunların sonuçları ve prognozları tartışılmıştır.

INTRODUCTION

Chondrosarcomas are malignant cartilaginous tumors and arise in bone or superimpose on benign cartilaginous tumors. These tumors are slow growing but locally aggressive malignancies, rerely encountered in head and neck region and they display a propensity for progressive spread and multiple recurrences that eventually lead death, if inadequately treated (1).

Although a lot of papers in medical literature have been published on this subject, most of them are case reports and anecdotal (2-10) and only a few papers have presented experiences of some large Institutions (1,11-17). In addition, there are broad discussions on the treatment of chondrosarcomas of the head and neck.

This article was written for the purpose of review of literature, concerning histolo-

* *Assistant Prof. of Gaziantep University, Faculty of Medicine, Department of Ear Nose and Throat Surgery - GAZİANTEP*

gic charasteristics, pathogenesis, grading, cinical features, management and prognosis of chondrosarcoma of the head and neck.

In this paper, literature data which was collected for a retrospective study that has planned in Institute of Laryngology and Otology (London) in which I have attended for three months between August 1991 to November 1991, has been presented.

PATHOGENESIS

There is considerable doubt about the origin of all cartilaginous tumors. The predisposition for chondrosarcoma of the anterior mandible and posterior maxilla has led some authors to postulate that the tumor arises in association with cartilaginous remnants of the nasal capsule and Meckel's cartilage respectively. On contrary, some other authors suggested that chondrosarcoma can arise de novo from osseos tissues without presence of cartilaginous rests (18).

The most frequent predisposing conditions include hereditary exostosis, Ollier's disease and Mafucci's syndrome. Previous intravenous thoratrast contrast use, Paget's disease of bone, chondromyxoid fibroma and previous irradiation represent less frequent associated conditions (1).

HISTOLOGIC FEATURES

Histologic spectrum of chondrosarcoma is ranged form well differentiated growth which may be difficult to differentiate from a benign cartilage tumor, to that of a high grade malignancy which has aggressive local behaviour and potential for metastasis (14). Evans et al. (15) divided chondrosarcomas into three grades based on their cellularity, mitotic rate and nuclear size (Table 1).

TABLE 1
CRITERIA FOR HISTOLOGIC GRADING OF CHONDROSARCOMA

GRADE	CRITERIA
I	Usually uniform and not densely cellular. Chondroid or myxochondroid matrix well developed. Rare to absent mitoses. Small, uniform nuclei often two or more in a single lacuna.
II	Less than 2 mitoses per 10 high power fields. Cluster density of nuclei, usually at periphary. Nuclei are larger and less uniform. Matrix less chondroid.
III	Two or more mitoses per 10 high power fields. Prominent cluster density, usually at periphery. Nuclei largest of the grades. Spindle cell forms and poorly developed matrix.

Differentiation of chondroma and chondrosarcoma can be very difficult, since they may appear identical grossly and can be very similar histologically. Many lesions

diagnosed as chondromas may actually be early or well-differentiated chondrosarcoma. For this reason, the diagnosis of chondroma should be treated with some skepticism, and multiple biopsy specimens should be taken, particularly, since variable degrees of differentiation within a tumor are common (5). Also, in chondrosarcoma, the differences from normal may be quite subtle and severety of these differences provides an indication of the propensity for aggressive growth of laryngeal cartilage neoplasm. These criteria are as follows: a) There are too many cells. b) The cells and nuclei are too irregular and pleomorphism is present. c) Nuclei stain too darkly. d) Large or giant cells with single, double or multiple nuclei are present (19).

CLINICAL FEATURES

Incidence:

Chondrosarcoma, also known as "chondrogenic sarcoma" (18,20) is the second most common primary malignant tumor of bone after osteosarcoma. In the head and neck, however, chondrosarcomas are not common malignancies and %10 of all chondrosarcomas occur in this region (1,4,14). On the other side, chondrosarcomas account for 10% to 20% of malignant primary bone tumors (1).

Locations:

The body areas most commonly involved by chondrosarcomas are the pelvis, sternum, ribs, clavicles, long bones of the lower limb, shoulder and scapula, long bones of the upper limb and then head and neck and foot respectively (1,21). In the head and neck, the most common sites of origin are paranasal sinuses, nasal cavity and septum, skull base, temporal bone, larynx and mandible (1,16,22).

Age and Sex:

Chondrosarcomas most occur in an age range 30-60 years (1,5,11-13,19,23). It is generally accepted that males are predominantly affected with male to female ratios ranging 1:1 to 10:1 (1). This ratio was 2.7:1 in the series of Neel et al. (12), and 1.8:1 in the series of Burkey et al. (1). On contrary, this ratio was 0.8:1 in Finn et al.'s series (11).

SOME SPECIAL LOCATIONS IN HEAD AND NECK

Larynx:

Until now, about 160 laryngeal chondrosarcoma cases were collected from the world literature (7). Chondrosarcoma of the larynx accounts for less than 1% of all malignant tumors of the larynx (8). About one-fifth of the cartilaginous neoplasms of the larynx are malignant (23) and these tumors have a marked male predominance (8) generally arising in those between

40 to60 years of age (19,23). The majority (70%) arise from the posterolateral aspect of the cricoid cartilage, followed by the thyroid (20%) and arytenoid cartilages (23).

According to Cantrell et al. (6), 78% of these tumors arise from cricoid cartilage, 15% arise from thyroid cartilage, 5% from arytenoids and 2% from vocal cord. Jacobs et al. (2) reported a case arising from epiglottis.

The clinical picture may be dominated by airway obstruction rather than hoarseness. Direct laryngoscopy reveals a smooth submucosal bulge, generally in the subglottis. X-rays will demonstrate a homogenous density, often flecked with calsification. Biopsy is usually unrewarding because of the firmness of the mass (23). In Mayo Clinic series (12), between 1910 to 1979, 33 patients were seen and most of them had hoarseness and dispnea, several had impaired vocal cord mobility, and one-third had a lump low in the neck. Of the 33 patients, 21 were grade I, 10 were grade II and two were benign chondroma. In 25 patients, radiologic examination yielded positive findings. Plain lateral cervical x-rays showed various forms of the calcification within the tumor, displacement or deformity of the airway and most commonly, a round, well circumscribed soft tissue mass. According to these authors, the most helpful radiologic studies were anteroposterior tomograms of the larynx which provided clear-cut localisation of the tumors.

Jaws and Facial Bones:

The maxilla, especially the anterior alveolar region is the site of occurence in approximately 60% of the cases. In the mandible, the coronoid process, condyles, premolar or molar regions and the symphisis account for the majority of the tumors (20). Average age of the patients with craniofacial chondrosarcoma is 32 years (24). In the facial bones, chondrosarcomas outnumber benign cartilage lesions in this region by a ratio of, at least, 2:1. Therefore, all cartilaginous tumors in the craniofacial region should be suspect and one must be wary of a diagnosis of chondroma arising in the facial bones (20). Radiologic evaluation is essential before rendering a microscopic diagnosis. Lesions larger than 6 to 10 cm. should be considered chondrosarcoma until proven otherwise (24).

Nose and Paranasal sinuses:

In the nose and paranasal sinuses, chondrosarcomas are twice as common as benign chondromas and the peak insidence for age is in the third to fifth decades of life (5). In the sphenoethmoidal area, the differentiation among chondrosarcoma, chondroma and chordoma may be very difficult (17). Fu and Perzin (16) reviewed a series of 256 non epithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx and 4% of these cas s were chondrosarcoma. Chondrosarcoma of the nose and paranasal sinuses occur with symptoms of intranasal swelling or a mass lesion, nasal obstruction and pain. Larger tumors may also cause cranial nerve deficits. The natural history of chondros rcoma is a slow progression of growth characterized by cartilaginous replacement of normal bone with subsequent myxomatous degeneration. Pulmonary metastasis is a late and unusual finding and metastatic spread to local lymph nodes is extremely rare (25).

MANAGEMENT

Treatment of chondrosarcoma of the head and neck has posed a challenge because of varying biological behaviour at different anatomic sites (11).

The management of chondrosarcoma has been primarily surgical and the extent of surgery modified by site and size of the tumor (1,11).

When there is involvement of the facial skeleton or mandible, complete resection with adequate margins is necessary and this is perhaps the most significant factor in such tumors. The level of differentiation is also a major factor with the higher grade malignancies being more likely to be unresectable (11).

Chondrosarcoma with neuroaxial involvement has a tendency for repeated local recurrences where complete primary resection cannot be done. Tumors at the base of the skull are likely to have extensive spread by the time of diagnosis and subtotal resection is often the only possible surgical treatment (11).

Chondrosarcoma of the larynx has long been regarded as a separate entity, because, in that location, the tumor is usually-differentiated with a slow course and very rare metastatic capability (1,6,11,12,13). The hallmark of management has been resection of tumor with partial laryngectomy when it is possible to preserve enough laryngeal skeleton to avoid stenosis. Total laryngectomy is indicated in cases with widespread involvement of the larynx or with recurrence in which there is not enough of the laryngeal structures remaining to maintain airway competence (1,6,11,12).

Although en bloc resection is the main therapy, in some patients conservative resection or surgical debulking may be more usuful with the preservation of some important structures. Unresectable lesions are not cured by other modalities but postoperative radiation therapy may be beneficial for controlling some of the such tumors (1,6).

The use of postoperative radiotherapy for high grade or extensive lesions has not routinely been reported. However, Harwood et al. (17) stated thad chondrosarcomas were sensitive to irradiation and potentially radiocurable. Clasically, described as a radioresistant tumor because of prolonged response time to irradiation, chondrosarcomas are not routinely offered postoperative adjuvant radiotherapy. However, the utility of postoperative radiotherapy in cases of unresectable disease or inadeqate surgical margins was suggested by series of chondrosarcoma at all body sites form some large centers (17,26).

In the series of Harwood et al. (17), 38 patients were treated with curative irradiation to, at least, 4000 cGy and none of them received complete surgical excision of the tumor. The 5 and 10 year actuarial survival rates were 41% and 36% respectively, With local control achieved in 47% of the cases. Suite et al. (26) reported a series of patients treated with dodes ranging from 6350 to 7500 cGy. These tumors were all unresectable cases of the cervical spine and skull base, and dise-

ase free survivals ranged from 4 to 74 months postirradiation. Based on the studies mentioned above, it has been suggested that radiotherapy be used both as primary treatment for unresectable chondrosarcomas and postoperatively in situations of inadequate surgical margins.

Little has been written about chemotherapy for chondrosarcoma. Chemotherapy may be used for palliation (11). Burkey et al. (1) reported that no tumoricidal effect was observed. On contrary, Finn et al. (11) reporting on three patients with extensive nasal chondrosarcoma treated with chemotherapy, noted partial responses in all patients and one survivor to 5 years, but no cures.

PROGNOSIS AND SURVIVE

The prognosis for chondrosarcoma is best judged by three factors: Tumor site, tumor grade and resectability, Tumors that arise in the larynx are generally believed to be more differentiated and less aggressive than chondrosarcomas that arise in other body sites (1). Of chondrosarcomas in the head and neck, nasopharyngeal and posterior nasal cavity tumors have the worst and laryngeal tumors have the best prognosis. This factor and resectability, are probably most important in determinig survival. If the tumor cannot be resected with adequate margins, the chance or differentiation of the tumor is also important; higher grades do less well than lower grades (16).

The local recurrence rate for chondrosarcoma of the head and neck is 85 percent, versus 15% for tumor elsewhere in the body (17). Death usually follows intracranial involvement, or less commonly, distant metastasis (1).

Five year survival for chondrosarcomas of grade I, II and III in all anatomic locations, are 90%, 81% and 43% respectively (15). Finn et al. (II) from MD Anderson Cancer Center showed an 81% survival in their series of 23 patients. Burkey et al. (1) reported overall survival of 70%. Fu and Perzin (16) reported a 69% survival in 10 patients with lesions in the nasal cavity, nasopharynx and paranasal sinuses. On the other side, according to Batsakis (20), the cure rate for chondrosarcoma of the jaws is 44% at 5 years. Verner et al. (27) reported 40-60 percent of the five years survival rate. Laryngeal chondrosarcomas usually have a good prognosis when adequately removed, with a cure rate of 75 to 85% (6,8,12).

These results are promising and and support en bloc surgical resection as the mainstay of therapy in head and neck chondrosarcomas. Conservative resection of low grade lesions in the temporal bone and larynx to preserve important structures, though did result in long term survival (1,6,13).

CONCLUSIONS

Chondrosarcomas are slow growing but locally aggressive malignancies with a propensity for progressive spread and multiple recurrences. The management of these tumors is mainly surgical and en bloc resection must be done whenever pos-

sible. However, if the tumor is unresectable, conservative resection or surgical debulking and then radiotherapy may help in local control of the disease. Chemotheerapy may offer for tumors with aggressive local spread or metastasis. The prognosis is depended to the anatomic site, grade and resectability of the tumor.

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