Chondrosarcoma of the nasal septum*

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SUMMARY

Chondrosarcoma of the nasal septum is a rather uncommon tumour. Therefore, only limited numbers of cases have been described in the literature. This paper reveals one more case observed at the ORL department of Dicle University. In the present case, by using a transnasal and transpalatal approach, total resection of the tumour was performed in conjunction with postoperative radiotherapy.

Key words: chondrosarcoma, nasal septum

INTRODUCTION

Chondrosarcoma is a malign cartilaginous tumour. It occurs most frequently in the long bones, pelvis and ribs, but rarely arises in the head and neck (Jones, 1973; Hornibrook et al., 1983). Ten percent of all cases occur in the maxillofacial region and the larynx, but it arises rarely in the nasal septum (Mc Coy et al., 1981). In another series, 80% arose from nasal structure (turbinates, septum), with only 20% found in the maxilla (Kragh et al., 1960).

CASE REPORT

The patient, a 25 years-old man, Mr. M.Ç., was admitted to ORL Physician at Medical School of Dicle University in 1997. He complained of having nose bleeding, swelling in the hard palate and of headaches. The nasal obstruction of the patient, which had started four months earlier, had increased in time. A swelling in the hard palate, that started two months earlier, had grown and developed into an ulceration. The patient was, therefore, referred to the ORL Clinic of Dicle University for further evaluation and operation.

During clinical examination, anterior rhinoscopy showed that both nasal cavities were filled with a hard mass that had a smooth mucosal surface. Mouth examination revealed a hard mass of about 3×3 cm with smooth mucosal surface at the left front of the hard palate.

A computed tomographic scan revealed soft tissue filling the left nasal cavity and extending into the nasopharynx (Figure 1). The tissue was characterized by calcified centers and the deposition of contrast agent was absent. The left cavity was enlarged. The nasal septum was replaced to the right and had eroded. Furthermore, Figure 2 shows a lacunar structure containing

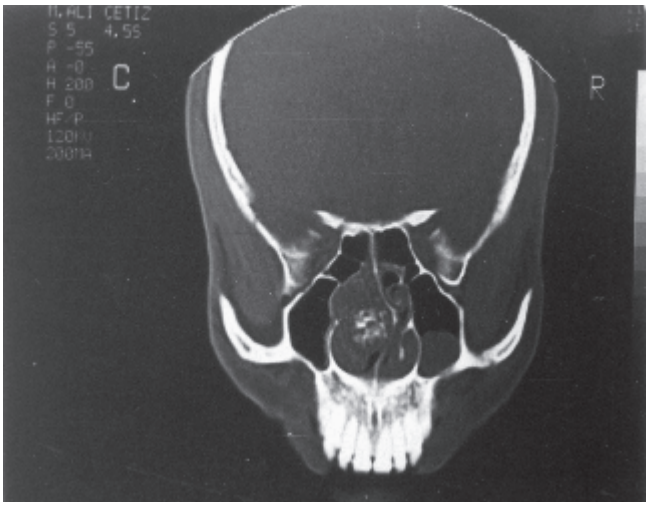

Figure 1. Coronal CT scan demonstrating a mass on nasal septum.

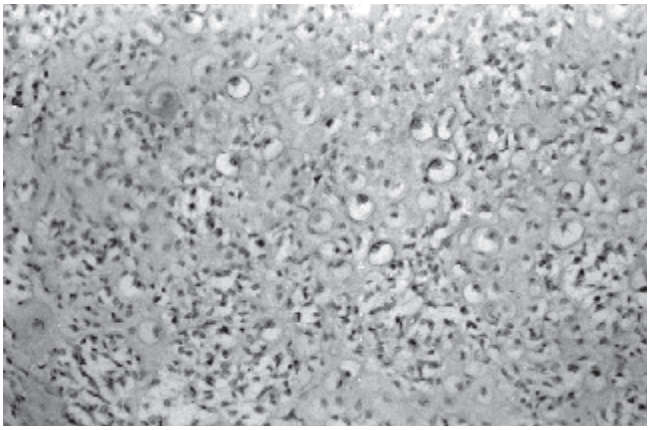

Figure 2. The nuclei are plump and hyperchromatic, and there are two or more nuclei per cell and two or more cells per lacuna.

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pleomorphic cells surrounded by a highly cellular sarcomatous area, which includes atypical cells with hyperchromatic nuclei (H.E. ×41). By using a transpalatal and transnasal approach the mass was excised, together with the hard palate, 2/3 of the posterior nasal septum and the inferior vomer.

During the postoperative period, the patient received 5000 cGy radiotherapy in total to the neck and 7000 cGy radiotherapy to the nasopharynx and the nasal cavity. At 16 months follow-up, the patient was well without evidence of recurrence or metastasis.

DISCUSSION

Characteristically, chondrosarcoma arises in the extremities, but some cases may be originated from the septal region (Jones, 1973; Hornibrook et al., 1983; Mc Coy et al., 1981). Most authorities agree that distinguishing chondrosarcoma from chondroma is histologically very difficult. However, some typical aspects such as hypercellularity, hyperchromatism, irregular multiple nuclei and an appearance convenient to sarcoma can be considered in favour of malignity (Randall et al., 1984). The histopathological examination of the specimens from our patient exhibited all these aspects.

The immunohistologic investigation may facilitate differentiation of mesenchymal cells (positive for vimentin) from epithelial cells (positive for cytokeratin, epithelial membrane antigen) (Roessner et al., 1984; Wang et al., 1993). The observation of a positive reaction to the S-100 protein and vimentin, and a negative reaction to cytokeratin and epithelial membrane antigen by the tumour cells of the chondrosarcoma, proves that this tumour has its origin in the cartilaginous tissue. Therefore, they have been used as important criteria to diagnose whether the tumour is chondrosarcoma.

Chondrosarcomas are classified into three main groups:

1- primary chondrosarcomas, arising from undifferentiated perichondrial cells, occur usually in younger patients. These are highly vascularized, extend into veins, and metastasize early.
2- secondary chondrosarcomas, arising from metamorphosed cells, occur in older patients.
3- mesenchymal chondrosarcomas, arising from primitive mesenchymal cells, occur predominantly in women and rarely in the head and neck (Myers and Thawly, 1979).

The criteria for the diagnosis of primary and secondary chondrosarcoma are 1- development from mature cartilage, 2- derivation from sarcomatous stroma (never osteoid tissue), and 3- cellular and nuclear irregularity (Lichenstein and Jaffe, 1942). The tumour in our case was present in the nasal perpendicular plate, in the septal cartilage and in the vomer. It was therefore considered to be primary chondrosarcoma. Nasal obstruction is the major symptom in 78% of the cases (Nishizawa et al., 1984). The main complaint of our case was also nasal obstruction. Wide surgical resection is the main treatment of choice for the chondrosarcomas of the head and neck (Hornibrook et al., 1983; Nishizawa et al., 1984). Although radiotherapy or chemotherapy alone is reported to be less effective, postoperative conventional high-dose radiotherapy and fast-neutron radiation are suggested as adjuvant treatments (Harwood et al., 1980).

Various surgical approaches such as antrectomy by a Weber-Ferguson incision, sublabial transnasal approach, the lateral rhinotomy approach and the craniofacial approach have been used to deal with chondrosarcomas (Arestsky et al., 1970). However, a standard approach should be a combination of approaches that provide the widest visualization of tumour and with the widest circumferential resection margins. The transpalatal combined with the transnasal approaches used in our case provide these conditions.

The factors affecting prognosis are the degree of tumour differentiation, adequacy of resection, location and the extent of the tumour (Harada et al., 1990; Beneck et al., 1984). The tumours were grouped to into Grade I, II and III on the basis of mitotoic rate, cellularity , and nuclear size. The five-year survival rates of Grades I, II, and III were 90%, 81%, and 43%, respectively. The overall five-year survival rate was 77%, and the 10-year survival rate was 67%. The initial surgical treatment (curettage or resection of all the involved bone) has an important role in local recurrence, but not survival (Evans et al., 1977). The nasopharynx, sphenoid sinus and skull base are the sites with the poorest prognosis (Hornibrook et al., 1983). No invasion in the posterior nasal cavity was a positive aspect for the present case.

It has been reported that the local recurrence rate increased about 25% after three years follow-up (Randall et al., 1984). Local recurrence developed in 93% of chondrosarcoma patients with Grade I and II who were initially treated by local excision or curettage, but in only 16% of those treated by resection (Evans et al., 1977). In our case, recurrence and metastasis had not occurred during the 16 months follow up.

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