Metastatic Nasal Chondrosarcoma

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SUMMARY

Chondrosarcomas are uncommon tumors which originates in the head and neck and accounts for approximately 10 to 15% of all cases. Most chondrosarcomas exhibit an indolent growth pattern. Nasoseptal tumors may mimic common sinonasal conditions, making early diagnosis difficult. We report a case of nasoseptal metastatic chondrosarcoma which was atypical, characterized by an aggressive growth pattern and widely disseminated.

KEY WORDS:
Nasal, Chondrosarcoma, Metastatic

INTRODUCTION

Chondrosarcomas are uncommon tumors and that which originates in the head and neck and account for approximately 10 to 15%[1] of all cases. Chondrosarcomas of the nasal septum are rare. Most chondrosarcomas exhibit an indolent growth pattern. Nasoseptal tumors may mimic common sinonasal conditions, making early diagnosis difficult. Yet despite late identification, disease is often localized, and skull base and cranial nerve involvement have been reported to occur in only 22% of cases. We report a rare case of nasoseptal metastatic chondrosarcoma which was atypical, characterized by an aggressive growth pattern and widely disseminated.

CASE REPORT

A 47 year old, Chinese man presented to ENT clinic in 25 May 2003 with complaints of right nasal stuffiness. Examination revealed a skin nodule obliterating the whole of right anterior nasal space. He was a known case of chondrosarcoma at level T4 and T5 diagnosed in 1998 for which subsequent excision was performed without any complications. Unfortunately he had a recurrence of chondrosarcoma at the same site in 2000 for which he underwent a second surgery, which led to post operative paraplegia with neurogenic bladder. Excision biopsy of the skin nodule from nasal septum, which was occupying the right nasal cavity, was performed under general anesthesia in year 2003.

The histopathological examination result was metastatic chondrosarcoma (Fig 1 & 2). Thereafter multiple swellings were recorded over the right parietal and left temporal scalp, nuchal region, right hypochondrium and left arm. He was readmitted again in September 2003 for right gluteal tumor, which bled profusely. On auscultation, there was decreased air entry over right middle and lower zones of the right lung with bilateral basal crepitations. Chest x-ray showed haziness over the right lung. Wide excision of the gluteal tumor was performed immediately and hemostasis secured. Unfortunately patient developed dyspneoa on 3rd postoperative day and his condition gradually deteriorated and patient expired on 6th postoperative day.

DISCUSSION

Most of the previously reported cases of nasoseptal chondrosarcomas occurred during the fourth and fifth decades of life. Men and women were equally affected. Chondrosarcoma is a malignant, slow growing, cartilaginous tumor most commonly found in the pelvis, ribs, and long bones. Approximately 5-10% is located in the head and neck, mostly in the maxilla and mandible. Nasal septal origin is rare. Based on their cells of origin, they can be classified as one of three types; primary, secondary, or mesenchymal. Primary chondrosarcomas arise from undifferentiated perichondrial cells and ooccurs usually in younger patients. It is highly vascularized and metastasizes early. Secondary chondrosarcomas originate in transformed cells from a central chondroma or cartilaginous exostosis and occurs in older patients. Mesenchymal chondrosarcomas develop in primitive mesenchymal cells[2] and has the poorest prognosis. Chondrosarcomas are graded histiologically according to their degree of cellularity, atypia, mitotic activity, nuclear size, and surrounding matrix composition. Grade I tumors have an ample chondroid matrix with scattered clusters of chondrocytes with near-normal nuclei and no mitotic figures. Grade II tumors display a higher degree of cellularity with a less-chondroid matrix, increased mitotic figures, multinucleation, and hyperchromatic vesicular nuclei. Grade III tumors are characterized by irregularly shaped chondrocytes in a myxoid matrix and increased nuclear pleomorphism. Imaging studies reveal characteristic findings. Computed tomography demonstrates a low-density matrix with speckled calcifications and bony erosion. CT scan demonstrates characteristic scattered ring-forming calcifications among a hypo dense matrix and often-bony erosions[3]. The ring like calcifications is related to the pattern of calcification inside the tumor and can be observed microscopically. MRI helps delineate the full extent of the soft tissue, typically demonstrating low intensity on T1-weighted imaging, high intensity on T2-weighted imaging, and heterogenous enhancement with gadolinium contrast. Radiographic differential diagnoses include chondroma, osteoblastoma, osteochondroma, meningioma, and fibro-osseous lesions. Chondrosarcomas are generally slowly growing malignancies. Their gradual progression allows for

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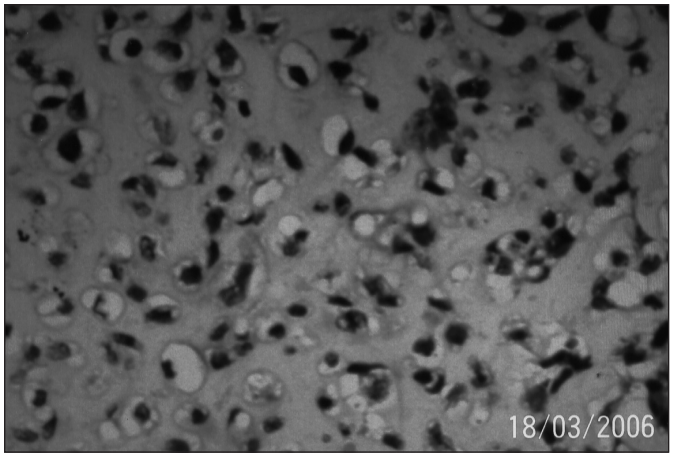

Fig. 1: There are malignant cells exhibiting pleomorphic and hyperchromatic nuclei and some of the cells are vacuolated. (H & E x 40).

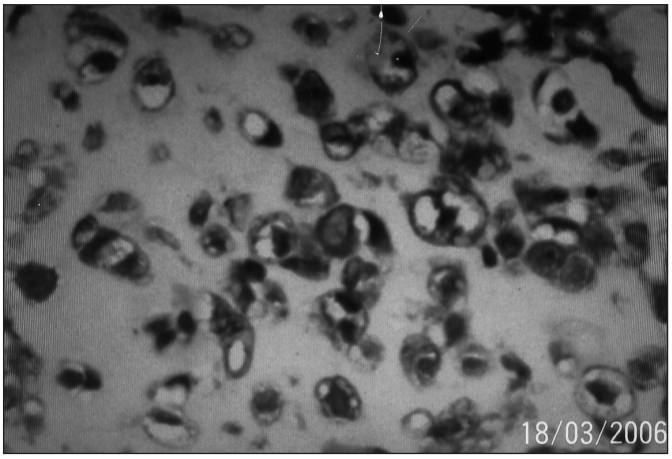

Fig. 2: The pleomorphic cells are positive for S100. (IP x 40).

relatively asymptomatic growth, which typically leads to a late diagnosis. This unfortunately can result in extensive loco-regional infiltration prior to diagnosis. In cases where the lesion breaches the cranial vault, the extent of involvement usually remains extramural and is limited to the anterior cranial fossa. The clinical presentation depends on the site of origin and the extent of local disease. Patients whose tumors involve the sinonasal area often present with nasal obstruction of one year's duration. Less common complaints include headache, epistaxis, anosmia, cranial nerve palsy, and various visual disturbances. Complete surgical resection is the treatment of choice[4]. Incomplete resections are associated with a 65% recurrence rate. Radiation and chemotherapy are reserved for recurrences and for cases of incomplete extirpation. Although total resection improves outcomes, surgical treatment may prove to be difficult in cases of extensive local disease. The overall 5-year survival rate is 44-81%[2]. Approximately 20% patients develop distant metastases, mostly to the lungs. Uncontrollable local recurrence and progression of disease are the usual causes of death. Although they are malignant tumors, chondrosarcomas often show features suggestive of a benign lesion. Often young surgeons may mistake them for benign lesions because of their well-circumscribed appearance and lack of periosteal reaction.

Secondary chondrosarcomas in the head and neck are very rare when compare with Primary tumour. Their indolent growth patterns mimic common sinonasal conditions making early diagnosis rather difficult. Hence, as an otolaryngologist, one should have metastatic chondrosarcoma as one of the many differential diagnosis especially with history of Primary chondrosarcoma

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