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Chondrosarcoma of the Nasal Septum

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ABSTRACT

A case of chondrosarcoma of the nasal septum is presented with the result of treatment. The patient was admitted for a growth in the nose of four years' duration. Fine needle aspiration for cytological examination was suggestive of squamous cell carcinoma. She was treated with lateral rhinotomy and wide excision followed by septorhinoplasty. Histological examination showed that the lesion was chondrosarcoma. The patient remained free of disease 26 months after surgery.

Keywords: chondrosarcoma, nasal septum, sinus neoplasm

INTRODUCTION

Chondrosarcoma of the head and neck region is a rarely encountered tumour. It has a tendency for progressive spread and multiple recurrences and hence is considered to have a poor prognosis[1]. Widest possible exposure and wide surgical excision are considered the treatment of choice. We describe here a case of chondrosarcoma of 4 years' duration arising from the nasal septum, together with the surgical approach and post-operative result.

CASE REPORT

BD, a 78-year-old Malay female, presented with a history of a mass arising from the nose of 4 years' duration. It was very slow-growing, painless and the patient reported occasional epistaxis. For two years prior to admission, the growth had completely blocked the nose resulting in absolute mouth breathing. The patient had an offensive foul-smelling discharge from the growth for one week prior to admission. There was no history of trauma, fever, loss of appetite or weight-loss. She had suffered a cerebrovascular accident four years previously with residual right sided hemiplegia. She gave no history of tuberculosis or diabetes mellitus. Clinical examination revealed a firm and non-tender intranasal mass involving both nasal cavities (Fig 1). A probe was passed all round the mass through both nasal cavities and there was no bleeding. Posterior rhinoscopy was carried out and results showed that the mass did not extend to the choanae or nasopharynx. Also, there were no palpable lymph nodes.

Investigations of the full blood count, ESR, liver and renal function tests, random blood sugar, chest X-ray and ECG were normal. Her VDRL and TPHA were found to be positive but on repetition, there was no increase in titre. Soft tissue X-ray of the face and computerised tomographic scan (Fig 2) revealed a well circumscribed mass with calcification. The hard palate and sinuses were normal. Although the clinical and radiological pictures were suggestive of a benign tumour, fine needle aspiration cytology however, suggested a malignant tumour, which was probably well-differentiated squamous cell carcinoma.

The patient was subjected to an extended lateral rhinotomy and wide excision of tumour with septorhinoplasty. The tumour was found to arise from the cartilaginous nasal septum alone. Since the frozen section reported the margins as free of malignancy, the surgical defect was repaired by a pedicled buccal mucosal flap from the cheek and bone graft for columellar support. The post-operative result is shown in Fig 3.

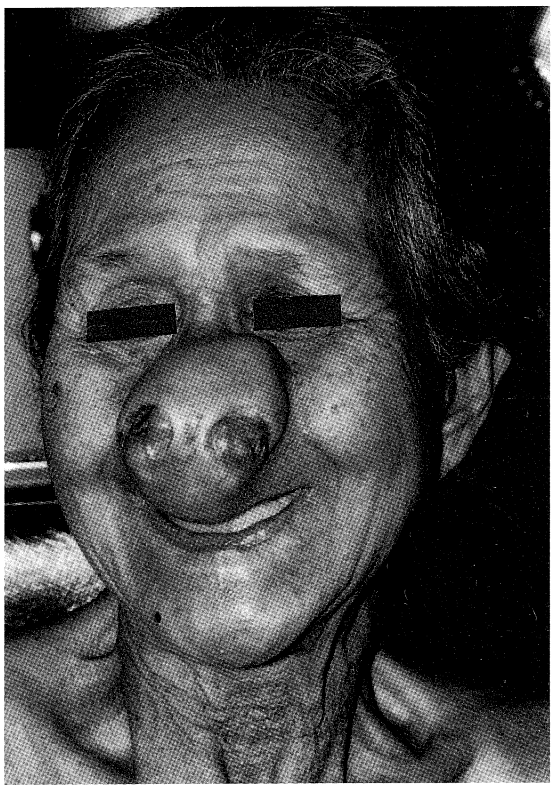

Fig 1 – Patient with tumour mass filling the nose.

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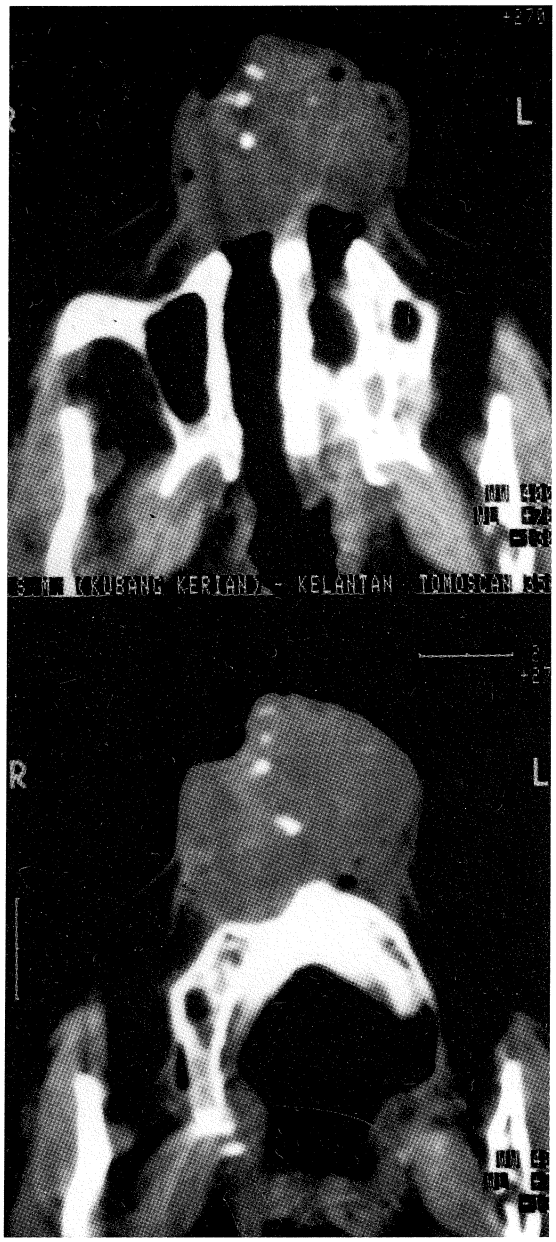

Fig 2 – CT scan axial cuts showing calcification of tumour matrix. There is no destruction of the hard palate.

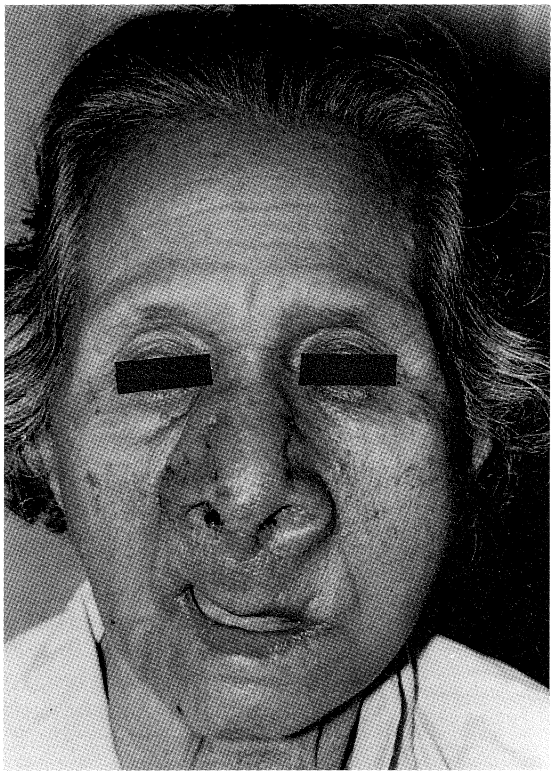

Fig 3 – Post-operative appearance after extended lateral rhinotomy, wide excision of tumour and septorhinoplasty.

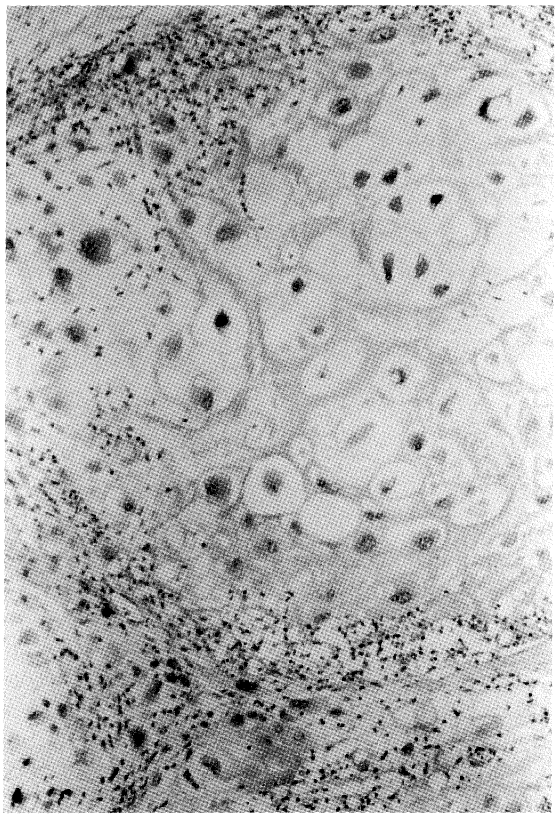

Fig 4 – Histology of the tumour showing tumour lobules made up of multinucleated malignant cartilaginous cells with nuclear hyperchromatism, pleomorphism and mitosis (H&E x 400).

Pathological findings

Upon examination, the consistency of the tumour was found to be circumscribed, grey white, lobulated and firm. The cut surface revealed grey-white lobules of varying sizes, with glistening hue and cartilaginous consistency. Microscopically, the tumour was made up of clusters of chondroid cells having pleomorphic, hyperchromatic nuclei with occasional mitosis. Bi- and multinucleated cells were frequently seen. The tumour cells had vacuolated cytoplasm but some were intensely eosinophilic with irregular cytoplasmic processes resembling spider cells (Fig 4). Spotty calcification was also noted from the examination. The tumour extended up to the nasal mucosa though the resected margins were spared. The tumour gave a stronger positive reaction for Alcian blue-PAS stain and immunohistochemical markers for myoglobulin and S-100 protein. A diagnosis of chondrosarcoma of moderate grade (grade II) of the nasal septum was made.

The patient has been on regular follow-up for 26 months till the time of this report without any recurrence of disease.

DISCUSSION

Neoplasms of the nose and paranasal sinuses constitute only 0.2% to 0. 8% of all neoplasms of the body. Of these, only 27% to 35% arise from the nasal walls[2]. Chondrosarcoma constitutes only 10% to 20% of all malignant primary bone tumours, of which only 10% arise from the head and neck areas[3]. The most common sites are the larynx, maxilla and skull base[1]. Chondrosarcomas constitute only 4% of non-epithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx[4]. Hence this malignancy is considered as one of the rarest. Only 28 cases have so far been reported from the nasal cavity and septum[5]. The highest incidence is among males in the fourth decade of life, making this case of a female in her eighth decade of life presenting with chondrosarcoma of the nasal septum an unusual one.

Due to their slow growing nature, chondrosarcomas tend to be large, multiloculated and sharply demarcated. Tumour matrix calcifications are almost always present[6]. Although the histological appearances of this tumour are generally characteristic, a differential diagnosis of chondroma, chordoma, and chondromyxoid fibroma should be considered. In our case, the tumour was made up of clusters of chondroid cells having pleomorphic hyperchromatic nuclei with occasional mitosis. Bi- and multinucleated cells were frequently seen. These features described as the cytological criteria for chondrosarcomas[7] are usually absent in chondroma except in periosteal chondroma and enchondroma. However, chondromas are not as cellular as enchondomatosis and never as cellular as central chondroma. The cells of solitary enchondroma are often larger than normal chondrocytes and may lie in well found capsulated lacunae; they occur singly or may be arranged in pairs or tetrads and though they vary in size and shape, they are mostly round and contain a single darkly stained nucleus[8].

Typical physaliferous cells suggestive of chordoma or a lobular arrangement of cells with crowding of cells at periphery typical of chondromyxoid fibroma are also absent, thus differentiating it from these tumours. The presence of spider shaped cells immuno-histochemical localisation of myoglobulin in the cytoplasm and a fairly circumscribed appearance may give an erroneous impression of a skeletal muscle tumour. Fine needle aspiration biopsy through an area of chondroid matrix may show pools of amorphous material that can be mistaken for keratin, thus leading to an erroneous diagnosis of squamous cell carcinoma as in this case. However, S-100 and Alcian blue-PAS stains will confirm the diagnosis. It is not unusual for a fine needle biopsy or even an excision biopsy to be unreliable since the specimen may not be representative of the lesion as areas of cellular atypism may be restricted only to certain parts of a well-differentiated chondrosarcoma[8].

Therefore, it is important to evaluate multiple fields to grade these tumours accurately[9]. Since it is described as a radio-resistant tumour due to its prolonged response time to radiation, the standard doctrine of treatment is wide "en bloc" resection. Hence these tumours are not routinely offered post-operative adjuvant radiation therapy[1]. However, adjuvant radiation therapy or chemotherapy may have to be utilised for residual disease[10]. Prognosis of chondrosarcoma depends on the site, grade and resectability of the tumour. The worst prognosis has been implicated in centrally occurring chondrosarcomas of the pelvis, trunk, proximal extremity and head and neck areas especially the nasal cavity and nasopharynx[1].

However, there are contradictory views regarding the behaviour of this tumour[11]. Since the grade of the tumour is an important prognostic factor[12], high-grade lesions should be treated aggressively. Patients with incomplete resections requiring further radiotherapy or chemotherapy have a bad prognosis[10]. Chondrosarcoma can also get implanted in an operative scar[13] or even along the tract of a needle biopsy[14]. The implanted cartilaginous cells survive even in the most hostile environments[15] making it essential for a meticulous clearance of this tumour. In the patient documented here, there has been no evidence of recurrence for the past 26 months.

CONCLUSION

Chondrosarcoma of the nasal septum is an extremely rare tumour with a bad prognosis. A case of chondrosarcoma of 4 years' duration in a female patient treated surgically without any adjuvant radiotherapy or chemotherapy, is described here. Despite the long-standing nature of this malignant tumour, a complete resection was possible with a good post-operative result after 26 months of surgery. The patient is currently on regular 3-monthly follow-up.

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