Chondrosarcoma of nose and paranasal sinus – a rare presentation

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Abstract Chondrosarcoma, the most malignant cartilageneous tumor, constitute only 4% of non-epithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx, making it one of the rarest malignancies.^{1,3} Here we present a case of Grade 1 chondrosarcoma involving nose, maxillary sinus, ethmoids and sphenoid sinus managed surgically by wide excision. 12 months follow-up revealed no recurrence or complication.

Keywords Chondrosarcoma · Nose · Paranasal sinus.

Introduction

Chondrosarcoma of nose and paranasal sinus region is a rarely encountered tumour constituting only 4% of non-epithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx [1, 3]. It has a tendency for progressive spread and multiple recurrences and hence is considered to have a poor prognosis. Widest possible exposure and wide surgical excision are considered the treatment of choice. We describe here a case of chondrosarcoma of nose and paranasal sinuses in a 35-year-old male, together with the surgical approach and post-operative result.

Case report

A 35-year-old man presented to ENT outdoor of Calcutta National Medical College with the complaint of progressive nasal obstruction for the last 5 to 6 months. Initially the obstruction was on the right side and gradually became bilateral for the last 2 months. He also complained of recurrent spontaneous bleeding from the right side for the last 2–3 months and slight nasal discharge for the same duration. There was no history of pain trauma, fever, loss of appetite or weight-loss.

On anterior rhinoscopy, a reddish globular mass was seen to be present in the posterior part of the left nasal cavity principally occupying the region of the middle meatus. Right nasal cavity revealed a pale looking mass in the posterior part of middle meatus. On Probe test, in left side the mass was found to be firm in consistency. In right side the mass was firm below the overlying polypoidal mucosa. Mass was bleeding slightly on touch and was non-tender. The probe could not be passed on either side of the mass in both nasal cavities. Posterior rhinoscopy revealed a red fleshy mass in the left choana. On local examination, there was no significant palatal bulging mal-occlusion of teeth, swelling of the cheek or any eye signs. No palpable lymph nodes were present.

Diagnostic nasal endoscopy was done which revealed a reddish globular mass occupying the left middle meatus region arising probably from the septum. Right nasal cavity revealed a pale mass covered with edematous mucosa.

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Fig. 1 Contrast enhanced axial CT scan showing large homogeneous mass with speckled calcification occupying both left and right nasal cavity with bulging of left lateral nasal wall.

Fig. 2 Contrast enhanced coronal CT scan showing involvement of sphenoid sinus with no bony erosion.

X-Ray PNS OM view showed a dense homogeneous opacity occupying both right and left nasal cavity with haziness of left maxillary antrum. CT Scan with contrast showed a large homogeneous mass with speckled calcification occupying both left and right nasal cavity extending to the left maxillary, ethmoid and sphenoid sinus (Fig.1, Fig.2) Left lateral nasal wall was partially eroded (Fig.3)

Operative procedure

The patient was operated under general anaesthesia with orotracheal intubation. The mass was approached by lateral rhinotomy incision. The mass was seen to be arising from the posterior part of nasal septum. The main bulk of the mass was in the left nasal cavity with extension into the left maxillary sinus, ethmoids and sphenoid sinus. A small part of the mass was also present in right nasal cavity in the region of middle meatus without any erosion of adjacent structures. The mass was meticulously removed in Toto with most of the cartilaginous septum leaving part of bony septum and columellar cartilage intact to maintain the integrity of the nasal dorsum. The mass was large (7×5cm) lobulated, pinkish

Fig. 3 Contrast enhanced coronal CT scan showing huge homogeneous mass with speckled calcification occupying both nasal cavities and ethmoids with erosion of lateral nasal wall and mass bulging into left maxillary antrum.

Fig. 4 Specimen of mass (7×5cm) removed after surgery.

Fig. 5 Histopathological slide of the specimen (H&E stain, 10×10 X).

Fig. 6 Histopathological slide of the specimen (H&E stain, 40×10 X).

white and firm in consistency (Fig.4). Cutting the tumor mass gave a gritty sensation. Cut surface revealed areas of focal calcification with some cartilaginous foci. Histology of the tumor revealed enlarged plump nuclei, multiple cells per lacunae, many binucleated cells, slight nuclear pleomorphism, along with intervening spaces of chondroid and myxoid changes which was consistent with the morphology of classical grade 1 chondrosarcoma (Fig 5, Fig 6).

Discussion

Neoplasm of the nose and paranasal sinuses constitute only 0.2% to 0.8% of all neoplasm of the body.¹ Chondrosarcoma, the most malignant cartilaginous tumor, constitutes only 10% to 20% of all malignant primary bone tumours, of which only

10% arise from the head and neck areas [2]. Chondrosarcoma constitute only 4% of non-epithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx [1, 3]. Hence this malignancy is considered as one of the rarest. The highest incidence is among males in the fourth decade of life as seen in our case. Chondrosarcoma can occur in both very young and old age groups, from 3 months to 76 years of age according to various literature. The mesenchymal subtype showed a tendency to occur at a younger age. It is very slow-growing painless tumor and the patient usually reports occasional nasal bleeding as seen in our case.

On radiological examination, plain skull radiography showed only bone destruction and calcifications. On CT Scan, these tumours present with a soft tissue mass expanding and destroying bone and typically (89%) showing areas of nodular or plaque-like calcification on CT as evident in our case. The magnetic resonance characteristics are more specific and when present with the typical CT features are diagnostic of chondrosarcoma. They combine high signal on T2-weighted sequences, with differential enhancement on post-Gadolinium T1-weighted scans. The contrast enhancement is seen at the periphery of the tumour and the central chondromatous core does not enhance [4].

The pathological classification of the subtypes grade I–III is based on differences in characteristics such as nuclear size, cellularity, mitotic rate, and frequency of lacunae with multiple nuclei. On gross examination, chondrosarcoma is a grayish-white, lobulated mass. It may have focal calcification, mucoid degeneration, or necrosis. Grade I chondrosarcoma is characterized by mild hypercellularity, large vesicular nuclei, and small nucleoli; the five-year survival rate is 90%. Grade II displays moderately increased cellularity with a slightly poorer 5-year prognosis (81% survival). Grade III is much rarer than I or II and carries a 5-year survival rate of 43%. The hallmarks include marked hypertrophy, pleomorphism, and the presence of giant tumor cells [5].

Although the histological appearances of this tumour are generally characteristic, a differential diagnosis of chondroma, chordoma, and chondromyxoid fibroma should be considered.

With the help of tumour markers such as vimentin, cytokeratin, and S-100, the chondrosarcoma can be differentiated from chordoma. Chordomas lack vimentin immunoreactivity and chondrosarcomas fail to express cytokeratin. S-100 protein expression is present in both.

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 [3]. However, adjuvant radiation therapy or chemotherapy may have to be utilized for residual disease [6]. 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 [3].

However, there are contradictory views regarding the behaviour of this tumour. Since the grade of the tumour is an important prognostic factor, high-grade lesions should be treated aggressively. Patients with incomplete resections requiring further radiotherapy or chemotherapy have a bad prognosis. Chondrosarcoma can also get implanted in an operative scar or even along the tract of a needle biopsy. The implanted cartilaginous cells survive even in the most hostile environments making it essential for a meticulous clearance of this tumour [6].

Distant metastases in chondrosarcoma are found in 10% of grade II and 71% of grade III tumours [5].

Radiotherapy is another treatment for these tumours in the direct vicinity of essential structures. The anatomical close relation between the tumour and these critical normal structures limits the dose that can be delivered with conventional radiation treatment. Charged particle radiotherapy combined with three dimensional treatment planning results in superior dose distribution that allows delivery of high tumour dose with acceptable dose to the normal tissues.

Conclusions

Chondrosarcoma of nose and paranasal sinus is a very rare tumor with malignant potential. Diagnosis is made on the basis of radiological and pathological features. Prognosis depends upon the grade of the tumor and location of primary tumor. The tumor being radio-resistant, widest possible exposure and wide surgical excision are considered the treatment of choice. Post operative follow-up is mandatory as the recurrent disease is more difficult to treat.

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