Endoscopic Resection of Primary Nasoseptal Chondrosarcoma

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SUMMARY

Chondrosarcoma of the nasal septum is an extremely rare malignant tumor. It accounts for only 10% to 20% of primary bone tumors, with approximately 10% found in the head and neck. A case is presented here to illustrate its presentation, evaluation and surgical treatment.

KEY WORDS:
Chondrosarcoma, Nasal septum, Endoscopic resection

INTRODUCTION

Nasal septum chondrosarcomas is an extremely rare tumor accounting for only 10% to 20% of primary bone tumors, with approximately 10% found in the head and neck[1]. Surgical resection is the primary modality of choice. Adjuvant therapy is reserved for residual or recurrent disease and palliation. Various surgical procedures had been described in the literature[1-4]. We describe one case whereby the tumor was successfully resected endoscopically.

CASE REPORT

A 15 year old Chinese boy presented to the ENT clinic in May 2007 with history of progressive nasal block and epistaxis for the past four months. Office nasal endoscopic examination revealed a large, firm midline mass occupying both sides of the nasal cavity. A subsequent biopsy was reported as chondrosarcoma. CT scan of the brain and orbits demonstrated a large, heterogenously enhancing mass occupying the nasal cavity and extending into the right sphenoid sinus and the ethmoid air cells. There was bony expansion and destruction of anterior wall of sphenoid sinus (Fig. 1a). However, the anterior cranial fossa and the skull base was intact (Fig. 1b). Chest X-Ray did not show any evidence of metastasis.

The lesion was completely resected endoscopically. Intra-operatively, a well-encapsulated expansile mass, cartilaginous in nature, was seen involving the posterior third of the nasal septum. It extended laterally in between the two middle turbinates. Superiorly, it extended up to the cribriform plate and anterior cranial fossa and posteriorly eroding into the anterior wall of the sphenoid sinus. There was no evidence of orbital or intra-cranial involvement. Intra-operative frozen sections confirmed negative margins. The histopathological diagnosis was confirmed as chondrosarcoma of nasal septum (Grade II).

A repeated biopsy of the sphenoid cavity two weeks later showed complete clearance of the tumour. The patent has been on our follow-up for one year and has shown no signs of recurrence. A repeat chest X-Ray also shows no signs of distant metastasis to the lungs.

Post-operative CT scan of the paranasal sinuses after one year (Figure 2) revealed no tumor recurrence. The posterior part of the nasal septum, the anterior and posterior ethmoid air cells, the post-medial walls of the maxillary sinuses and the anterior face of the sphenoid have been surgically removed.

DISCUSSION

Head and neck chondrosarcomas commonly occurs in the larynx, maxilla and skull base[1]. Therefore lesions arising from the nasal septum is extremely rare. The most common presenting symptom is nasal obstruction[2]. Our patient presented with this primary symptom.

Chondrosarcomas are classified into three main types: (1) primary chondrosarcomas, arising from undifferentiated perichondrial cells and occurs in younger patients. It is highly vascularized and metastasizes early, (2) secondary chondrosarcoma, arises from altered cells either in a central chondroma or cartilaginous exostosis. It occurs in older patients, (3) mesenchymal chondrosarcomas, arises from primitive mesenchymal cells and has the poorest prognosis[2].

The histopathology of chondrosarcoma is well established[2]. It is divided into 3 grades based on the degree of cellularity, mitotic activity, and nuclear size and atypia. Grade 1 tumors have an abundant chondroid matrix with clusters of chondrocytes with normal or slightly irregular nuclei, rare nuclei, and absent mitoses. Rare binucleate cells may be present. Grade 2 lesions demonstrate more cellularity, less matrix, the presence of mitoses, enlarged nuclei and a higher number of binucleate cells. Grade 3 tumors exhibit high cellularity, prominent nuclear pleomorphism, prominent mitotic figures and stellate or irregulary shaped chondrocytes[2]. Our patient demonstrated a lesion of Grade 2 variety.

Imaging is crucial before undertaking any surgical intervention. CT scan in cases of chondrosarcoma of the nasal septum demonstrates a hypodense matrix with scattered small calcifications with occasionally large or ring-like calcifications. Erosion of the septum and surrounding structures bony structures are common[2]. On the other hand,

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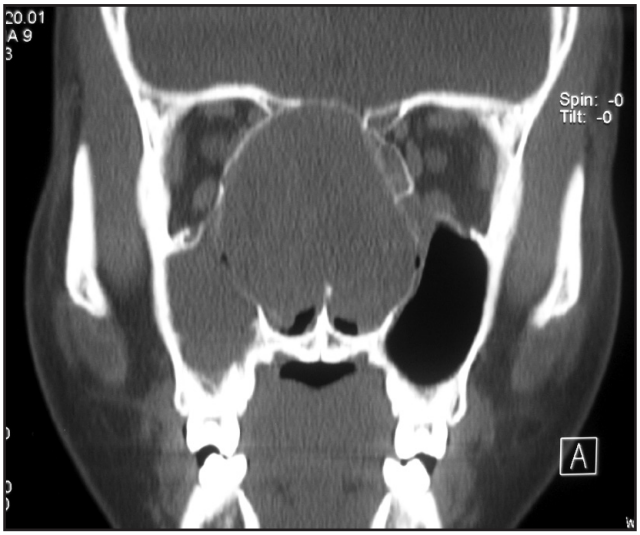

Fig. 1: Pre-op CT scan (Coronal View, Bone Window) of paranasal sinuses showing the huge well-encapsulated mass occupying the midline of the nasal cavity. The nasal septum is entirely engulfed by the tumor.

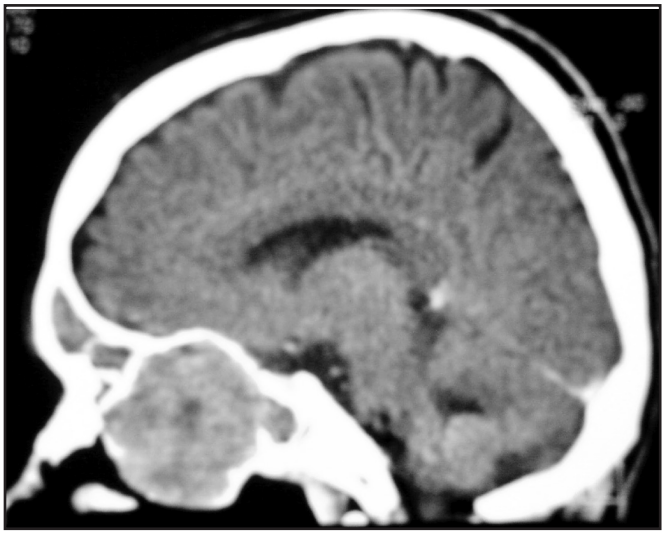

Fig. 2: Pre-op CT Scan (Sagittal view) showing the mass extending into the sphenoid cavity. The skull base is intact with no intracranial extension.

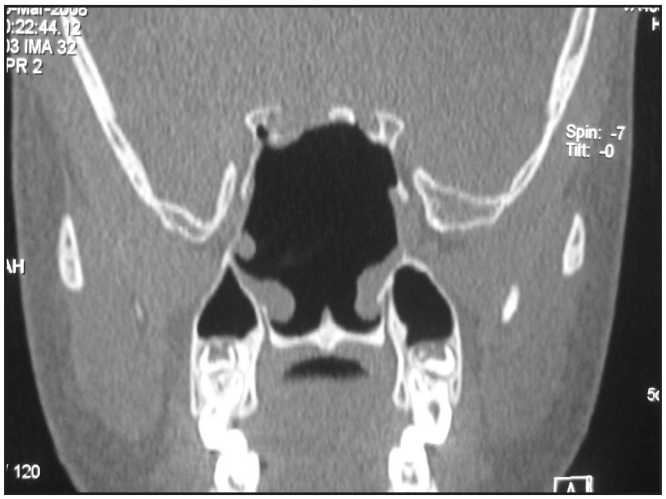

Fig. 3: Post-operative CT scan (Coronal view) after one year showing complete tumor clearance.

MRI shows images of low intensity on T1 weighting, high intensity on T2 weighting, and inhomogenous type of enhancement on T1 weighting on gadolinium[2]. The radiographic differential diagnosis of skull base neoplasms in the nasal cavity includes chondroma, osteoblastoma, meningioma, osteosarcoma and chondroid chordoma should be borne in mind.

Surgery is the definitive treatment of choice. The goal of surgery is histologically clear margins, as the reported recurrence rate is as high as 65% with positive margins[2]. Various surgical approaches has been described in the literature: Weber-Ferguson, lateral rhinotomy, sublabial transnasal, Le Forte I downfracture, anterior craniofacial approach and transnasal excision[1-4]. Endoscopic excision has been reported in the literature[1,4]. Lesions limited to the nasal cavity, without evidence of intracranial, skull base or orbital extension are suitable for endoscopic resection[1,4].

Endoscopic surgery provides superior magnification, illumination, no facial scarring, superior cosmesis and angled visualization facilitating complete endonasal tumor resection. Tumor and adjacent mucosa and bone can be removed with powered microdebriders and diamond burr. In contrast to many external approaches, preservation of normal sinonasal physiologic function and mucociliary clearance patterns is achieved with the endoscopic approach. Potential morbidity with the more open and extensive approaches can also be spared. Hospital stay is minimized and post-operative care is similar to any other patient undergoing endoscopic sinus surgery.

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