CLINICAL VIGNETTE

A Rare Case of Sinonasal Chondroblastoma

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Introduction

Chondroblastoma (CBT) is a rare tumor that originates within the epiphysis of the long bones and accounts for less than 1% of all primary bone tumors.[1-2] This neoplasm is a solitary lesion that most commonly involves the tibia, distal femur and proximal humerus. The chondroblast is thought to be the cell of origin. The chondroblasts are accompanied by secondary elements such as mature cartilage, giant cells and calcification.[3-4] This disease usually occurs in men and is more commonly diagnosed during second decade of life.[5] The radiographic features of CBT are that of well-demarcated, lytic lesions and averaging 3-6 cm.[6]

CBT of the craniofacial region is a very rare. In 2015, only 20 cases of craniofacial CBT had been reported. The craniofacial CBTs often emanate from the squamous portion of the temporal bone and present in the third decade of life.[7]

History of Present Illness

A 58-year-old female presented with nearly one year history of sinus congestion, loud snoring, obstructive sleep apnea symptoms, headaches, hyposmia and fatigue. The most significant symptoms were unremitting sinus congestion and recurrent sinus infections. The physical examination was unremarkable. For many months the patient was treated with courses of antibiotics and decongestants.

The patient was eventually referred to ENT for further evaluation. A limited sinus computed tomography (CT) revealed large nasal cavity/nasal septal lesion measuring 5.2 x 3.5 x 5.8 cm. The lesion was causing local mass effect and dehiscence and erosions in the cranial fossa/cribriform plate. A PET-CT revealed a FDG-avid mass in the sinonasal cavity, consistent with malignancy. There was no evidence of metastatic lesions. The MRI of the face showed nasal cavity mass measuring 3.4 x 4.5 x 3.8 cm. The mass eroded through the cribriform plate and ethmoid roof, and transversed the olfactory nerves.

A biopsy was performed which revealed malignant tumor that could not be further defined. She then underwent an endoscopic endonasal resection of anterior skull base tumor, bilaterally maxillary antrostomy, ethmoidectomy, sphenoidotomy, endoscopic CSF leak repair with Alloderm and fascia lata lumbar drain placement. During surgery, the large mass was noted to be centered in the nasal septum, obliterating the ethmoid sinuses and with intracranial/dural extension. A gross total resection was achieved endonasally. Frozen margins were negative for tumor. The final pathology revealed Chondroblstoma (CBT) with atypical features.

The postoperative course was unremarkable with improvement in sinus complaints. Anosmia/hyposmia persisted. Postoperative MRI revealed no evidence of residual mass. She did not receive adjuvant therapy. Six months after surgery, she remained in complete remission.

Discussion

CBT of the craniofacial region is rare and comprises approximately 7% of all cases.[8] The patients are typically older.[5] Patients may present with various signs and symptoms depending on the exact anatomy. They may have hearing loss, otalgia, cranial neuropathy, sinus congestion/pain, diplopia, headaches and facial swelling.[9,10] The differential of such presentation is broad but possibilities include but are not limited to plasmacytoma, chondrosarcoma, chondroma or chondromyxoid fibroma.

The exact cell origin of CBT is unknown.[11] However, the required histopathologic features include the chondroblasts (mononuclear cells), osteoclastic-like giant cells and a chondromyxoid stroma.[10,12] High mitotic rates are uncommon and most display low proliferative rates.[13]

The CBTs are generally benign and the prognosis depends on the local recurrence rate as they rarely metastasize.[4,14] Therefore, metastatic workups are not the norm. Local recurrences are as high as 29% in the flat bone lesions and 11% of the long bone lesions. The skull and temporal bone regions have the highest recurrence rates of 50%. The dismal recurrence rates seen in the skull may be a reflection of the regional challenges of a complete excision.[4]

Surgery is the primary modality of therapy for CBT. Complete and en bloc surgical resection is mandatory[15,16], as simple curettage has been associated with high recurrence rates.[17] There is no role for systemic chemotherapy and or adjuvant radiation. Of note, are reports of radiation-induced chondrosarcoma.[15] Radiation therapy is used for recurrent disease, incomplete resections or for patients who are poor surgical candidates.[15,18]

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