CHONDROSARCOMA OF SPHENOETHMOID REGION: REPORT OF A CASE

SFENOETMOİD BÖLGE KONDROSARKOMU : VAKA SUNUMU

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SUMMARY : Chondrosarcoma is a malignant tumor that has a slow growth with late metastasis. It is rarely seen in the head and neck and even less frequently in the paranasal sinuses. Congenital and acquired factors are involved in etiology. Chondrosarcoma arising from the sphenoethmoid region gives rise to symptoms especially by invasion of vital structures. Headache and visual disturbances are the most common of these symptoms. Computed tomography (CT) and magnetic resonance imaging (MRI) aid in diagnosis. Treatment of chondrosarcoma is radical surgical excision but radiotherapy and chemotherapy are additional treatment modalites in selected cases. A case of chondrosarcoma of sphenoethmoid region is presented in this study.

Key Words: *Chondrosarcoma, Sphenoethmoid Region, Paranasal Sinus.*

ÖZET : *Kondrosarkom malign tümör olmasına karşın yavaş büyür ve geç metastaz yapar. Baş boyun bölgesinde özellikle paranazal sinüslerde çok az rastlanır. Etyolojisinde konjenital ve akkiz faktörler mevcuttur. Sfenoetmoid bölgeden köken alan kondrosarkomlar vital yapılara invazyon sonucu semptomlar oluşturur. Baş ağrısı ve görme bozukluğu en sık yol açtığı semptomlardır. Tanısında öncelikli olarak bilgisayarlı tomografi ve magnetik rezonans görüntüleme kullanılır. Tedavisi radikal cerrahi eksizyondur. Radyoterapi ve kemoterapi ancak radikal cerrahi eksizyonun yapılamadığı durumlarda ek tedavi olarak uygulanır. Bu çalışmada sfenoetmoid bölgeden köken alan kondrosarkom vakası sunulmuştur.*

Key Words: *Kondrosarkom, Sfenoetmoid Bölge, Paranazal Sinüs.*

INTRODUCTION

Chondrosarcoma is the second most common primary malignant tumor of bone tissue following osteosarcoma. It usually derives from cartilagenous tissues and enchondral bone tissues (1, 2). It arises most commonly from the pelvic bones and long bones with a decreasing occurrence in the proximal femur, proximal humerus, distal femur and ribs (2). Twelve percent of chondrosarcoma arises in head and neck sites (2, 3). It rarely originates in the nasal cavity and paranasal sinuses (4, 5). The peak incidence is in the third to sixth decades of life (3). Histologic type of tumor varies with age and sex distribution. Female to male ratio of chondrosarcomas of the nasal cavity and the paranasal sinuses is 3 to 2 (2). The natural process of chondrosarcoma is slow growth with a late metastasis and relatively good prognosis (1, 3).

CASE REPORT

A 63-year-old male patient was referred to our department with complaints of headache which increased continuously over 3 months and referred to the right eye. He had no visual disturbance. His past medical history was otherwise unremarkable. Physical examination revealed a large mass in the right nasal cavity. On nasal endoscopy, the mass was observed to be

covered with mucosa extending from the right sphenoethmoid region to the nasopharynx. The remainder of the physical examination was normal. On CT scans an irregular mass arosing in the sphenoethmoid region which destroyed the medial wall of the right maxilla, anterior skull base, medial wall of the right orbit and posterosuperior septum was demonstrated (Fig. 1). The optic nerve was intact. Magnetic resonance imaging showed a 4,5 x 7 x 6 cm hypointense mass in T1-weighted sequences, and hyperintense in T2-weighted sequences. Contrast enhancement was present at the periphery of the mass (Fig. 2). Multiple punch biopsies were

Fig. 1: Axial CT scan of chondrosarcoma with nodular calcification (arrow) in the sphenoethmoid region showing bone destruction.

Fig. 2: Post-gadolinium T1-weighted coronal MRI sequence showing bone destruction, contrast enhancement at the periphery of the mass (arrowheads).

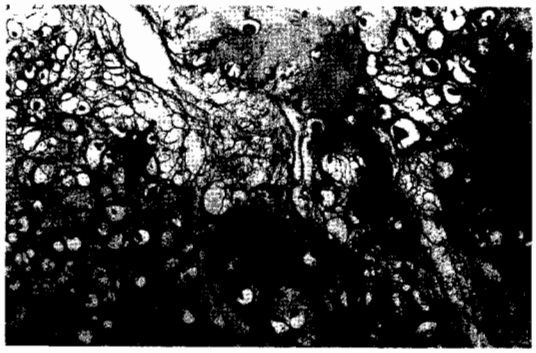

Fig. 3: Grade I chondrosarcoma; hypercellularity, hyperchromatism, nuclear pleomorphism within well differentiated chondroid stroma. HE x 200.

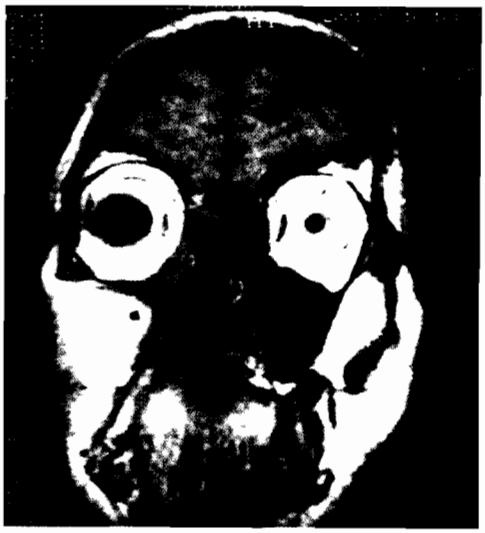

Fig. 4: Post-gadolinium T1-weighted coronal MRI sequence demonstarting tumor-free cavity in postoperative 7th month.

obtained from the mass. Histopathological examination revealed grade I chondrosarcoma (Fig. 3).

The location, size and extention of the tumor determined our surgical approach. The tumor was planned to be resected by total maxillectomy and total sphenoethmoidectomy with the option of craniofacial resection in cooperation with neurosurgeons. During the operation it was possible to resect the entire mass with total maxillectomy and total sphenoethmoidectomy. Oncologic safety was confirmed with multiple negative frozen sections. A small dural defect and

2x2 cm bone defect in the ethmoid roof was repaired by a rotating flap from the nasal mucosa of the septum and tissue glue. Maxillectomy defect was reconstructed by obturator prothesis in the third postoperative week.

His postoperative course was uneventful. During follow-up, multiple biopsies were obtained from the cavity and were negative in the postoperative month. Radiologic control was assessed one year later and there was no sign of recurrence on MRI (Fig. 4).

DISCUSSION

Various etiologic factors have been held responsible for chondrosarcoma. These include malignant transformation in the ossification centers or cartilage remnants of the enchondral bones; differentiation of primitive mesenchymal cell to malignant chondroblasts; although debated by many authors, physical and chemical trauma including radiation, radioactive isotope, beryllium, zirconium and lucite (2, 6, 7). Additionally, a possibility of hereditary tendency of chondrosarcoma has been emphasized by Scajowicz and Bessone (8). According to Lund, sphenoid and ethmoid bones originate enchondrally from Bertini's ossicle (concha Bertini) (9). Reino states that sphenoethmoid chondrosarcomas arise either from malignant transformation in the ossification centers of enchondral bones or from the remnants of Bertini ossicle in the postpartum period (7).

Chondrosarcomas are classified as primary and secondary. Primary chondrosarcomas derive from cartilage, bone and soft tissue whereas secondary chondrosarcomas derive from benign (enchondroma, osteochondroma) or malignant tumors (2). The probability of development of chondrosarcoma is high in multiple hereditary exostosis, Mafucci's syndrome, Ollier's disease, Paget's disease of bone, fibrous dysplasia, chondromyxoid fibroma, osteosarcoma, fibrosarcoma, Ewing's sarcoma, melanoma and leukemia (2, 3, 10).

Symptoms of tumors which arise from the nasal cavity and paranasal sinuses occur due to involvement of vital structures, and size and extension of the mass (11). Vital structures neighboring the sphenoidethmoid region are dura mater, olfactory bulb and olfactory mucosa, pituitary gland, internal carotid artery, optic nerve, optic chiasm, cranial nerves III, IV, V1, V2, VI, sphenopalatine ganglion and artery, anterior and posterior ethmoid arteries, cavernous sinus, and pterygoid canal and its nerve (11, 12). Loss of vision and headache are cited as the first symptoms of sphenoethmoid chondrosarcoma (4, 5). Depending on the size and involvement of other structures diplopia, facial numbness, nasal obstruction, epistaxis, hoarseness, anosmia, facial pain, propitosis, hemiparesis, cervical lympadenopathy and CSF rhinorrhea may also be encountered (11). In our case, the first symptoms were progressive headache and retroorbital pain. The reason for this pain was thought to be involvement of the sphenopalatine nerve (11). Although the tumor was of considerable size, the patient did not have visual disturbance or neurologic symptoms.

To aid in diagnosis, CT and MRI provide valuable information. On CT scans, nodular or plaque-like calcification, lobulation and destruction of bone are compatible with chondrosarcoma. Contrast enhancement at the periphery of the mass on post-gadolinium T1-weighted seqeuence and very low or no enhancement at the center of the tumor is pathognomonic for chondrosarcoma on MRI. This feature on MRI is explained by excess vascularity and the cellularity at the periphery and central avascular zone of the tumor (13, 14). MRI is also important in follow-up. Small recurrent tumors that are missed in the routine nasal examination can be detected with MRI (14). Angiography can be used to demostrate the relation of the tumor with vascular structures (4).

In order to confirm diagnosis, biopsy should be performed. Currently chondrosarcomas are classified into 5 subtypes. Conventional chondrosarcoma (also referred to as chondrosarcoma) is the most common type and has the better prognosis (2). The other histologic subtypes are clear cell (malignant chondroblastoma), myxoid, mesenchymal and dedifferentiated variants. The differentiation of chondrosarcoma is classified as grade I (well differentiated), grade II (moderately differentiated) and grade III (poorly differentiated) (3, 5). Conventional chondrosarcoma is usually well differentiated and its prognosis is relatively better. Grade III chondrosarcomas are biologically more

aggressive and their local recurrence and metastasis are more frequent. Consequently, the prognosis is worse (3).

The main treatment method of chondrosarcomas is surgical excision. Depending on the location and extension of the tumor, surgical options include maxillectomy, ethmoidectomy, sphenoethmoidectomy and craniofacial resection. Wide resection should be performed since chondrosarcomas are pseudocapsular tumors. In order to determine microscopic residue, frozen sections should be obtained. Complete removal of the tumor is often curative. Chondrosarcomas are not radiosensitive, thus radiotherapy is not a primary method of treatment. Radiotherapy can be used in cases where the tumor cannot be removed completely. Chemotherapy has not been found to be benefical in chondrosarcomas (14, 15). Chemotherapy can be useful in selected cases such as chondrosarcoma associated with osteosarcoma of Ewing's sarcoma (2).

Prognosis of chondrosarcomas depends on location, size, histologic subtype, differentiation and resectabilitiy. Since removal of tumors from the posterior nasal cavity, sphenoid and nasopharynx is difficult, prognosis of chondrosarcomas arising from these sites are worse than those originating from the anterior nasal cavity and maxillary bone. Local recurrences of tumors that cannot be completely removed are more frequent (5). Survival rates of chondrosarcomas located in the head and neck are between 44-81% and their prognosis is worse than those arising from other regions (3).

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