CASE REPORT Libri Oncol., Vol. 32 (2004), N^{o} 1 – 3, 45 – 48

CERVICAL VERTEBRAL CHORDOMA – A CASE REPORT

ALEN SEKELJ, DAVORIN ĐANIĆ and IRENA PIRKL

Department of ENT and Head and Neck Surgery,
Dr. Josip Benčević General Hospital, Slavonski Brod, Croatia

Summary

Chordoma is a very rare primary tumor of bone and the only malignant tumor arising from the remains of the notochord (chorda dorsalis) upon regression of this embryogenic structure. Chordoma is typically localized along the midline, and classified into craniocervical, vertebral and sacrococcygeal groups according to its localization. Chordoma is characterized by slow growth, late clinical presentation, destruction of the adjacent vital structures, and high rate of metastasizing, mostly to the bone, liver and brain. A case of cervical vertebral body chordoma with retropharyngeal and cervical expansion is presented as a rare manifestation of the head and neck chordoma. Definitive diagnosis was made by tumor histopathology and immunohistochemistry.

KEY WORDS: *chordoma, cervical vertebra*

HORDOM TRUPA VRATNOG KRALJEŠKA: PRIKAZ SLUČAJA

Sažetak

Hordomi su vrlo rijetki primarni tumori kosti i jedini su maligni tumori koji nastaju iz ostataka notokorda (korde dorzalis) nakon regresije ove embriogene strukture. Karakterizira ih smještaj u medijalnoj liniji, a prema lokalizaciji se dijele u tri skupine: kraniocervikalne, vertebralne i sakrokokcigealne. Hordomi sporo rastu, kasno se klinički prezentiraju, vrlo često razaraju okolne vitalne strukture. Učestalost metastaziranja je velika, najčešće u kosti, pluća, jetru i mozak. Autori predstavljaju slučaj hordoma trupa vratnog kralješka sa širenjem u retrofarinks i vrat, do današnjih dana jednu od rijetkih prezentacija hordoma glave i vrata. Konačna dijagnoza se postavi patohistološkom i imunohistokemijskom analizom tumora.

KLJUČNE RIJEČI: *hordom, vratna kralježnica*

INTRODUCTION

Chordoma is a very rare primary malignant tumor of bone that arises from the remains of the notochord (chorda dorsalis), axial skeleton of a young embryo continuing in strip-like fashion from the middle of the cranial fossa to the end of the tail bone (coccyx). The incidence of this tumor ranges from 1% to 4% of primary malignant bone tumors (1, 2). Together with craniopharyngeal and colloid cystoma of the third ventricle, these tumors account for 3%-4% of the central nervous system tumors. Chordomas most commonly occur in advanced age, and are extremely rare in children. These tumors primarily develop in middle- to old-age groups, with an average age at onset of 46-58 years (3). In children, this tumor is associated with a wider array of symptoms, atypical histologic findings, tumor cell aggressiveness, wide cellularity spectrum on morphology, and high mortality rate (4, 5). Men are more frequently affected than women, with a 2:1 male

to female ratio (2). According to localization, chordoma is classified into three main groups: craniocervical (spheno-occipital), vertebral, and sacrococcygeal chordoma, with the respective incidence of 11%-37%, 10%-22%, and 50%-65% (2, 6). Embryologic studies in the head and neck region have revealed seven potential sites of its origin, the most common being dorsal aspect of the sella turcica, clivus, and nasopharyngeal region. Jallo et al. have proposed a new classification of chordoma based on its relation to dura mater and bone involvement into: type I, osseous extradural; type II, extraosseous extradural; type III, osseous intradural; and type IV, extraosseous intradural (2). A prepontine tumor localization is associated with headache, hemiparesis and tetraparesis, hyperreflexion, mydriasis, paresis and paralysis of cranial nerves III-XII, and cerebral signs. In case of intrasellar tumor localization, acromegaly, myxedema and bitemporal hemianopia are described, whereas suprasellar tumor localization is usually associated with headache accompanied by apathy, depressed mental activity, homonymous hemianopia and emotional alterations. Tumor expansion into the foramen magnum region always leads to paraparesis, hyperesthesia and coma. Vertebral C5-TH2 chordomas are restricted to the symptoms of upper extremity paresthesia and pathologic reflexes. Chordomas metastasize to long bone, lymphatic tissue, lung, liver, brain and skin, at a rate of 10% to 48% (3,7). Grossly, the tumor is of a milky-white color, smooth surface and mucinous appearance, lobulated and soft, occasionally with areas of hemorrhage. Externally, the tumor may show a cartilaginous appearance. Its similarity with chondrosarcoma, pleomorphic adenoma, mucinous adenocarcinoma, myxopapillary ependymoma and ecchondrosis physaliphora as well as with chondroid chordoma, a variant of chordoma containing chondromatous elements (8), accounting for as many as 14% (9), or according to some authors for 5%-35% (3) of all chordomas, has been emphasized. The diagnosis of this rare malignant neoplasm primarily relies on objective ENT, neurologic, neurosurgical and ophthalmologic examinations, computed tomography (CT) and nuclear magnetic resonance (NMR) findings, cytologic biopsy, and histopathology with immunohistochemistry.

CASE REPORT

A 66-year-old woman was admitted for clinical management of a tumor on the left side of her neck. For two years, she had suffered frequent headaches and hypernasality, with a slowly growing swelling on the left side of her neck present for the last nine months. The patient reported more severe headaches and pain around her left ear during the last two months. There were no vision, breathing, phonation or swallowing impairments. Oropharyngoscopy revealed a submucous tumor on the posterior wall of the pharynx, descending from the epipharynx along the posterior and left lateral wall of the hypopharynx to the level of the epiglottis (Fig. 1). The tumor pushed the lateral wall of the hypopharynx to the left piriform sinus. The endolaryngeal finding was normal. On rhinoscopy, the tumor

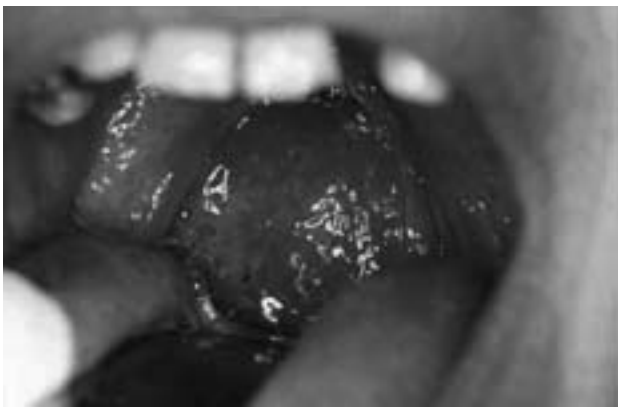

Figure 1. Oropharyngealy view – the submucous phryngeal tumor

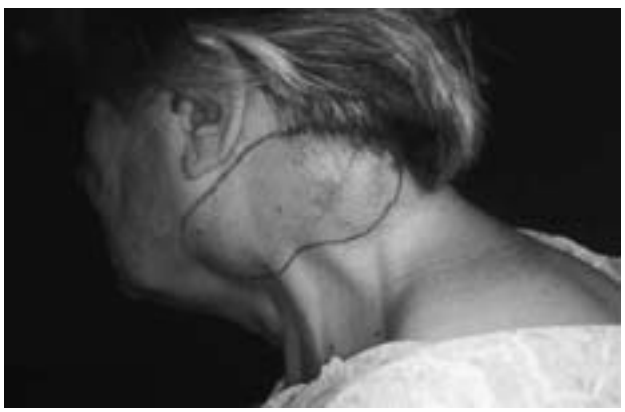

Figure 2. Multilobular and fixed left sided painless mass in the region II of the neck.

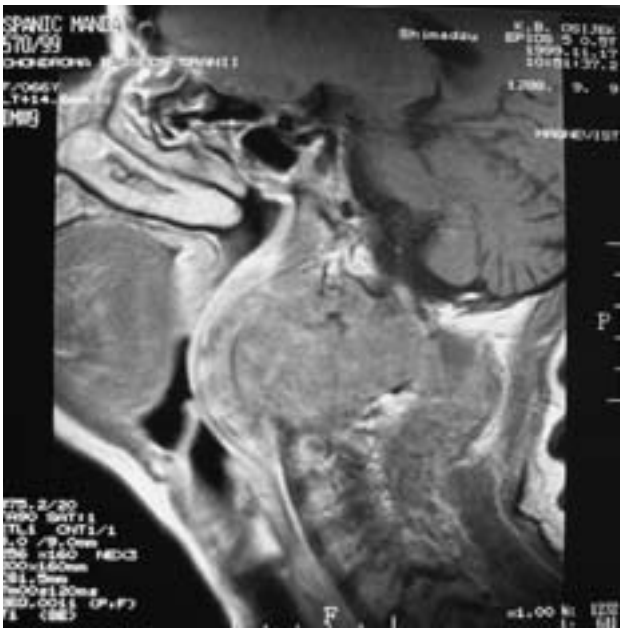

Figure 3. Parasagittal magnetic resonance image reveals inhomogenous paravertebral mass arising from the C2 vertebral body.

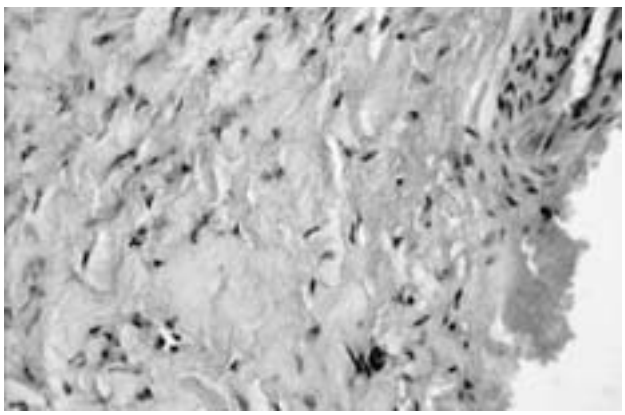

Figure 4. Section of tumor shows physaliferous cells with vacuolated, bubbly cytoplasm characteristic of chordoma. (Hematoxylin and eosin stain; original magnification x 200).

was found to have caused complete choanal obstruction on the left and partial obstruction on the right. The otoscopy finding was normal on the right, while revealing tympanum retraction on the left. A painless, sessile, multilobular growth was palpable in the left cervical region II, reaching the mastoid area posteriorly and the upper edge of the thyroid inferiorly (Fig. 2). Audiology showed conductive hearing loss in the left ear. The neurologic finding indicated no lateralization. Aspiration biopsy with tumor cytology revealed a myxotamous stroma with clusters of malignant cells. CT scan and magnetic resonance imaging (MRI) indicated a tumorous growth originating in the neck midline, at the level of C2 vertebral body. The tumor expanded to the left part of the epipharynx, completely filling the oropharyngeal lumen, and infiltrating the cervical musculature on the left to the subcutis, and the prevertebral and paravertebral musculature dorsally. Caudally, the tumor reached the lower edge of the cricoid cartilage, with overt infiltration of the vascular structures on the left side of the neck with lymph node involvement. The body and partially the arch of C2 vertebra, and the body of C3 vertebra were destroyed by the tumor, with spinal medulla compression at the level of the C2 vertebral body and along the entire C3 vertebra. Intracranially, no tumor lesion was observed (Fig. 3). Tumor tissue was partially excised by transoral access. Histologically, tumor tissue was composed of large cells with vacuolized cytoplasm, occasional mitoses (physaliphorous cells), and myxoid stroma (Fig. 4). On immunohistochemistry, the cells showed positive reaction to cytokeratin, EMA, S-100 protein, vimentin and GFAP, confirming the diagnosis of chordoma.

Radiotherapy at a dose of 70 Gy was prescribed. A 20% reduction of the tumor mass was recorded one year after the treatment. Then, tumor regrowth was observed, to cause death of the patient 40 months after the diagnosis.

DISCUSSION

Chordomas account for 0.2% of all nasopharyngeal tumors. The remains of the notochord are always found along the midline, occupying the medial area of the spheno-occipital synchondrosis in the head and neck region (8). Chordoma is extremely rare in the regions of frontal sinus, mandible and orbit, arising from the remains of the ectopic notochord tissue (10). The accurate diagnosis of chordoma is reached quite late, because of the slow tumor growth and noncharacteristic symptoms. Our patient had a type I tumor according to Jallo et al., with atypical symptomatology. Occasional headaches and rhinolalia did not pose major discomforts for

some two years. Although of large dimensions in the epipharynx and oropharynx, expanding to the left side of the neck, the tumor caused no neurologic events, swallowing or speech difficulties, or epistaxis. Thus, the patient presented exclusively for examination of the edema on the left side of her neck growing for the past 9 months.

The rate of survival of chordoma patients is poor, especially in the female population. The lowest rate is recorded in the group of postmenopausal women, therefore an association between sex hormones and growth of chordoma has been suggested (11). The 5-year survival for chordoma is 20% with a median of 4.1 years (1).

Complete surgical excision of tumor is the treatment of choice for small tumors localized on the head and neck. Large chordomas induce compression and involvement of the adjacent neurovascular structures, thus precluding complete tumor excision in most cases. Radical excision is significantly more difficult to perform in type I and III lesions or tumors with bone involvement.

Chordoma belongs to the group of radiosensitive tumors. If nonresectable, as in our patient, radiotherapy at a dose of 70 Gy or more is applied. Yet, chemotherapy is used as adjuvant treatment in case of poorly differentiated metastatic chordoma (12).

REFERENCES

1. Gladstone HB, Bailet JW, Rowland JP. Chordoma of the oropharynx: an unusual presentation and review of literature. Otolaryngol Head Neck Surg 1998;118: 104-7.
2. Jallo J, Nathan D, Beirbrauer K, Faber E. Chordoma: a case report. Surg Neurol 1997;48:46-8.
3. Khairi S, Ewend MG. Chordoma. Curr Treat Options Neurol 2002;4:167-73.
4. McMaster ML, Goldstein AM, Bromley CM, Ishibe N, Parry DM. Chordoma: incidence and survival patterns in the United States, 1973-1995. Cancer Causes Control 2001;12:1-11.
5. Sibley RK, Day DL, Dehner LP, *et al.* Metastasizing chordoma in early childhood: a pathological and immunohistochemical study with review of the literature. Pediatr Pathol 1987;7:287-301.
6. Nishigaya K, Kaneko M, Ohashi Y, Nukui H. Intradural retroclival chordoma without bone involvement: no tumor regrowth 5 years after operation. N Neurosurg 1998;88:764-8.
7. Peramezza C, Cellini A, Berardi P, *et al.* Chordoma with multiple skin metastases. Dermatology 1993;186:266-8.
8. Rossiello R, Ferrara G, Varricchio A, Baldi A, Motta S, Motta G. Chondroid chordoma of the lateral skull base. ORL J Otorhinolaryngol Relat Spec 2001;63: 114-8.
9. Horn KD, Fowler JC, Carrau R, Barnes EL, Rao UNM. Cytokeratin immunophenotyping of an unusual cervical vertebral chordoma with extensive chondroid foci and perilaryngeal recurrence: a case report with review of the literature. Am J Otolaryngol 2001;22: 428-34.
10. Moshari A, Bloom EE, McLean IW, Buckwalter NR. Ectopic chordoma with orbital invasion. Am J Ophthalmol 2001;131:400-1.
11. O'Connell MB, Renard LG, Liebsch NJ, *et al.* Base of skull chordoma: a correlative study of histologic and clinical features of 62 cases. Cancer 1994;74:2261-7.
12. Fleming GF, Heimann PS, Stephens JK, *et al.* Dedifferentiated chordoma: response to aggressive chemotherapy in two cases. Cancer 1993;72:714-8.

Received for publication: June 6, 2004

Author's address: Alen Sekelj, M.D., Department of Otorhinolaryngology and Head and Neck Surgery, General Hospital – Slavonski Brod, Slavonski Brod – Croatia,
E – mail: asekelj@net.hr and Alen.Sekelj@bolnicasb.hr