Clinical and Histopathological Findings of Chordomas: a Case Report

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INTRODUCTION

Chordomas are malignant tumors arising from the residual of embryonic notochord. They were first described by Lushka in 1856.1,2 These tumors are localized along the spine from the skull base to the sacrum. Chordomas are often seen in the sacrococcygeal region and clivus. Although these tumors are congenital, they are manifested in 4th and 6th decades. They are seen in men two times more commonly than women.3,4 They are accounted for 0.2% of nasopharyngeal tumors and 6% of all skull base tumors.5 Chordomas usually grow slowly and show local invasion. They have low malignancy potential, but are locally aggressive. These rare tumors are manifested by symptoms that vary according to the location of the tumor. They emerge with pain when localized in the sacrococcyx and with clinical symptoms due to cranial nerve compression when localized in the skull base. The most frequent involvement is seen in the 6th cranial nerve. The treatment is surgical excision. However, it is not possible to remove these tumors completely in many patients, due to their adjacency to important structures and invasive nature. Therefore, radiotherapy is needed after surgery.

CASE REPORT

CLINICAL ANALYSIS

A 78-year-old male patient was admitted to our clinic with the complaint of increasing hearing loss. In the examination both tympanums were dull and mildly retracted. In the endoscopic nasopharyngeal examination performed upon these findings a polypoid mass with a smooth surface, increased vascularization and a diameter of about 4 cm was detected in the midline (Fig. 1). Hearing test and contrast
enhanced nasopharyngeal MRI were ordered for the patient. In the hearing test, pure tone audiometry was measured as 66 dB in the right ear and 76 dB in the left ear, and bilateral mixed type hearing loss was detected. In the tympanometric examination, both ears were obtained as type B. Then, punch biopsy was performed from the nasopharynx.

Written informed consent was obtained by the patient.

MRI ANALYSIS

A tissue mass of 33×23 mm in the axial plane and 40×21 mm in the sagittal plane with a lobulated contour and which appeared to be tortuoused to the air column and showed a marked contrast enhancement in the postcontrast series was observed in the nasopharynx-globus localization (Fig. 2).

HISTOPATHOLOGICAL ANALYSIS

The material in the macroscopic examination was in the form of gray-white tissue. It was observed in the microscopic examination that the epithelial cells (physaliferous cells) whose small nucleoli can be distinguished in the myxoid zone and have oval round nuclei and wide eosinophilic vacuole cytoplasms created strata and cords in the lobular pattern (Fig. 3a). In the immunohistochemical study,

Figure 1. Endoscopic nasal view of the mass.

Figure 2. MRI view of the mass.

Figure 3a. Epitheloid cells with oval, round nuclei and large eosinophilic vacuole cytoplasms, which create strata and cords in the myxoid zone are observed (H&E, x200).

Figure 3b. Strong positive staining with Pan Cytokeratin immunohistochemical stain is observed (Pan Cytokeratin, x200).
these cells were positively stained with keratin, S100 and vimentin, and negatively stained with chromogranin (Figs 3b, 3c, 3d, 3e). Moreover, the histochemical study revealed positive staining of the myxoid ground with Alcian blue stain (Fig. 3f). The case was diagnosed as chordoma on the basis of these findings.

**DISCUSSION**

Chordomas are slowly growing, rare malignant bone tumors. Often, they reach large dimensions when they are diagnosed, and it is usually not possible to achieve complete cure. They are frequently located in the sacrococcygeal region (50%), at the skull base (30%), in the cervical region (10%), and in the thoracolumbar vertebrae (5%).4,6 Thirty to forty percent of the skull base chordomas originate from the clivus.3,7,8

Chordomas in the skull base usually grow in the clivus and may cause cranial nerve paralysis. Chordomas in the cella are often confused with pituitary tumors and manifest with hormonal disorders. Headache occurs in 75% of patients.9 Impaired vision and nasal obstruction are other symptoms. They rarely cause nosebleeds and intracranial hemorrhage.10,11 The patient we presented had symptoms of hearing loss and chronic serous otitis as clinical findings. We found no other case of chordoma with these findings reported in the available literature.

Diagnosis is usually established using plain radiography, computed tomography (CT) and MRI. But the definitive diagnosis is made by biopsy. CT and MRI acquired in the coronal and sagittal planes
show intra- and extracranial spreads of the lesion. In chordoma, destruction is radiologically observed in the bones involved along the median line. Half of the chordomas radiologically have calcification. In carotid or vertebral angiography, chordomas may be manifested as an avascular mass, localized in the midline of the clival region, pushing the basilar artery posteriorly and anteriorly. PET and CT are studies with high sensitivity that are used in the detection of metastases and follow-up of recurrence, respectively.

Chordomas are histopathologically divided into three subgroups: conventional chordomas, chondroid chordomas and dedifferentiated chordomas. Chondroid chordomas occur at younger ages and have better prognosis, and are frequently confused with chondrosarcomas. Dedifferentiated chordomas are very similar to high grade sarcomas and have a poor prognosis. Conventional chordomas are known to be well-circumscribed and to exhibit a lobular pattern. Histologically, epitheloid cells (physaliferous cells) with oval round nuclei and large eosinophilic vacuole cytoplasms, which create strata and cords in the myxoid zone, are observed. Although these tumors are often confused with nasopharynx carcinomas, salivary gland carcinomas and chondrosarcomas, the differential diagnosis is set through positive staining of the tumoral tissue with pancytokeratin, S100 and vimentin in immunohistochemical examination.

Surgery forms basis of the treatment of chordomas. Co-administration of surgery and radiotherapy in the treatment prolongs the life span. However, the cords may be radioresistant. Some studies have suggested that chordomas are radioresistant, and therefore the role of radiotherapy in their treatment is unclear. Nonetheless, the standard treatment of chordomas remains as complete resection and postoperative radiotherapy. More aggressive resections can be performed with technological advances in recent years (such as endoscopic surgery, navigation, neurophysiologic monitoring). In addition, radiotherapy performed with direct proton applications, carbon ions and modulation of stereotactic techniques helps to administer higher doses to the tumor site. Gamma-knife beam-surgery is an effective treatment for small chordomas. No effect of chemotherapy protocols tried on chordomas was observed so far. The prognosis of the chordomas is generally poor. Patients’ age, gender, histopathologic features, history of previous surgery and radiotherapy, the association of tumor with important structures, and the resection width are among the factors affecting the prognosis. Occipitocervical chordomas give earlier findings and are diagnosed earlier than the other localizations. Aggressive resection of skull base chordomas is challenging because of their proximity to important nerves, vessels and tissues and these chordomas cause serious complications.

Studies have shown that these tumors are more aggressive in patients over 40 years of age and in children under 5 years of age. In another study, small-sized tumors detected in children were found to have a longer survival time, but a higher risk of distant metastasis. Life of the untreated chordomas is 28 months from the time of diagnosis. Whereas the mean life span in treated cases is 3.6-6.6 years. The rate of metastasis in clival chordomas has been reported as 10-40%. They usually are manifested in late stages of the disease, and many cases are lost before metastases are identified. The cause of death is local recurrence associated with intracranial extension.

Tumoral formations such as chondrosarcoma, meningioma, invasive pituitary adenomas, plasmocytomas, metastatic tumors, sphenoid sinus carcinomas, nasopharyngeal carcinomas, lymphoma and rhabdomyosarcoma (in pediatric cases) should be considered in the differential diagnosis. Chordomas are commonly confused with chondrosarcomas. The chordomas are located in the midline of the clivus, whereas the chondrosarcomas are paramedian localized, and there are studies in the literature reporting chondrosarcomas seen in the petrous bone. However, there is no reliable method to distinguish chordomas from chondroid subtypes or chondrosarcomas that have different prognosis.

CONCLUSION

Skull base chordomas are rare malignant tumors for which surgery and radiotherapy used in a multidisciplinary way and that require long-term follow-up. We suggest that nasopharyngeal examination is a definite necessity in patients presenting with complaints of serous otitis media at an advanced age and that chordomas should be kept in mind among diagnoses.

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Клинические и гистопатологические находки хордомы: клинический случай

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Хордомы являются редким видом злокачественных опухолей, которые происходят из остаточного эмбрионального нотохорда. Эти опухоли наблюдаются по протяжению позвоночника и имеют локальное, агрессивное развитие. Хордомы у основания черепа часто встречаются в области затылка (затылочно-шейной кости). Эти опухоли, как правило, чрезмерно расширены при диагностике. Они локально инвазивны и редко развиваются отдаленные метастазы. Эти хордомы не могут быть полностью удалены из-за их локализации. Поскольку они находятся на продвинутой стадии при диагностике и находятся рядом с важными структурами, они относятся к опухолям с высокой степенью заболеваемости и смертности. При их лечении применяются хирургия и / или лучевая терапия.