

Synchronous Intrasellar and Cervical Chondroid Chordoma: A Case Report and Review of the Literature

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Abstract

We report on a patient who presented in ENT clinic with diplopia accompanied by headache and vomiting. Imaging studies revealed a large expansive heterogeneous mass encroaching on the intrasellar region and cervical spine. Post-intravenous contrast administration revealed a lesion with high enhancement and involvement of the sphenoidal sinus with retroclival extension. Biopsy confirmed the presence of a chondroid chordoma. Treatment included radiation therapy and the patient is currently receiving palliative and supportive care.

Keywords: Tumor; Chondroid; Chordoma; Head; Neck; Radiotherapy; Magnetic resonance imaging; Computed tomography

Introduction

Chordomas (CDS) are malignancies that arise from cellular remnants of the embryonic notochord [1]. The ratio of males to females with chordoma is approximately 1:1, between the third and fifth decades of life [2]. In the head and neck, these tumors are locally aggressive but metastasis to the lymph nodes of the neck is rare. Imaging studies are essential for preoperative staging and surgical planning [2]. The treatment of choice is complete surgical resection and adjuvant radiotherapy [1].

Case Report

A 68-year-old female with medical history of arterial hyper-

tension and type II diabetes mellitus presented to the ENT clinic complaining of diplopia of 4 months duration, headache, nausea and vomiting. Ophthalmic evaluation determined diplopia for six cranial nerve palsy. Endocrinologist evaluation was normal. A magnetic resonance imaging (MRI) was performed revealing an aggressive, expansive and destructive solid mass on the intrasellar region with involvement of sphenoidal sinus and retroclival extension with high enhancement and central hypointense areas following intravenous contrast administration suggestive of calcified deposits. In addition, an expansive and destructive process was observed in cervical spine at C2 level (Fig. 1). Computed tomography (CT) of head, neck and chest was performed, showing expansive and heterodense process in skull base and cervical spine with no evidence of cervical lymphadenopathies and the thorax was normal (Fig. 2).

A biopsy was performed under general anesthesia revealing a chondroid chordoma (CHCD). The immunohistochemistry tests were positive for protein S-100, cytokeratin AE1/AE3 and epithelial membrane antigen (EMA), and negative for glial fibrillary acidic protein (GFAP) (Fig. 3). The head and neck tumor board (HNTB) reviewed the case, where it was staged as a T4N0M0 tumor and determined out of surgical scope and recommended radiotherapy and palliative care.

Conventional palliative radiotherapy was performed with a Varian 21 iX linear accelerator, with a total dose of 36 Gy, administered in 3 Gy doses during a 2.5-week period (Fig. 4). At the end of the treatment, a neck MRI examination was performed revealing increased size of both tumors (Fig. 5). The patient was referred to the pain control and palliative care program.

Discussion

CDS are lesions of the middle line with low and intermediate grade of malignancy that arise from remnants of embryonic notochord in the craniovertebral axis, representing only 1-5.2% of all malignancies [1]. They are most commonly found in the sacrum bone (29-60%), skull base (25-36%) and cervical spine (15-34%) [3-7].

CDS in the head and neck are uncommon, accounting for less than 0.1% of all cases of the skull base malignancies. They are characterized as slow growing in adult patients, with equal distribution between males and females, with mean ages ranging from 30 to 50 years [2].

CHCD was described first time in 1968 like a biphasic

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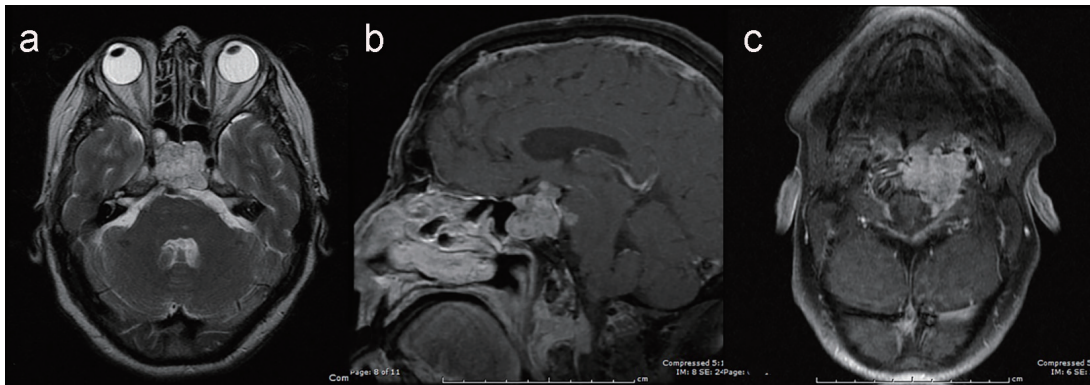


Figure 1. Magnetic resonance imaging (MRI) of solid expansive infraselar and cervical process with high enhancement post-intravenous injection of paramagnetic contrast. (a) T2 sequence axial view. (b) T1 fat sat and gadolinium sagittal view. (c) T1 fast sat and gadolinium axial view.

neoplasm with cartilaginous and chordal tissues in different proportions [8]. Finally, it was denominated CHCD in 1973 [5] and at present, there are less of 200 reported cases [2]. Most common locations of CHCD in the head and neck include clival region, sphenoparietal bone and sellar/parasellar region [2]. Usually the clinical findings are unspecific and depending on the location and include headache, diplopia, decrease vision, blurred vision, cranial nerve palsy (six), neck pain, nausea and vomiting [9].

Histopathologically, CHCD contains a significant amount of hyaline cartilaginous matrix in addition to the chordomatous component and are positive for protein S-100 in immunohistochemical study [10]. The histologic type is a consistent predictor of prognosis and its importance is illustrated in the American Joint Committee on Cancer (AJCC) staging system.

Since CHCD is frequently associated with soft tissue invasion and bone destruction, CT and MRI are essential for pre-operative staging and surgical planning in patients with these tumors [11]. CT scan findings include single hypodense and heterogeneous well-circumscribed mass with tumor calcifications. In late stage disease, the primary lesion may penetrate the cortical plate causing cortical expansion, destruction and extension into adjacent soft tissue. In MRI, in general, the

CHCD appears hypointense on T1-weighted, hyperintense on T2-weighted images with inhomogeneous enhancement after intravenous gadolinium administration [2].

Complete resection is the most effective primary treatment and may include soft and hard tissues with 82% survival rate, 22.6-60.3% of morbidity and 2-7.8% of mortality [9, 12-16]. The most common complications are neurologic deficits (13.9%), cerebrospinal fluid fistula and meningitis [12]. Positive margins and residual tumor is a powerful predictor of local recurrence [1]. Neck dissection is not indicated due to the low incidence of cervical node metastasis [1]. Adjuvant radiation therapy, with doses of 65 - 70 Gy, is recommended after surgery with positive margins and regional lymph node dissection for nodal metastases [17, 18]. Proton beam therapy is the gold standard for chordomas [19]. This improves the survival rates and decreases rate of recurrences to a 25% [1, 19, 20]. Chemotherapy is not effective [9, 20, 21].

The rate of local recurrence is about 68% [22]. The rate of local and distant metastases is around 5-29% [1]. The 5-year survival rate for CHCD ranges from 51% to 82.5%, according to the histological grade of malignancy and surgical margins, and the 5-year survival rates free of disease range from 33% to 65% [1, 9, 13-16, 21, 23-27]. Strict clinical and imaging (CT)

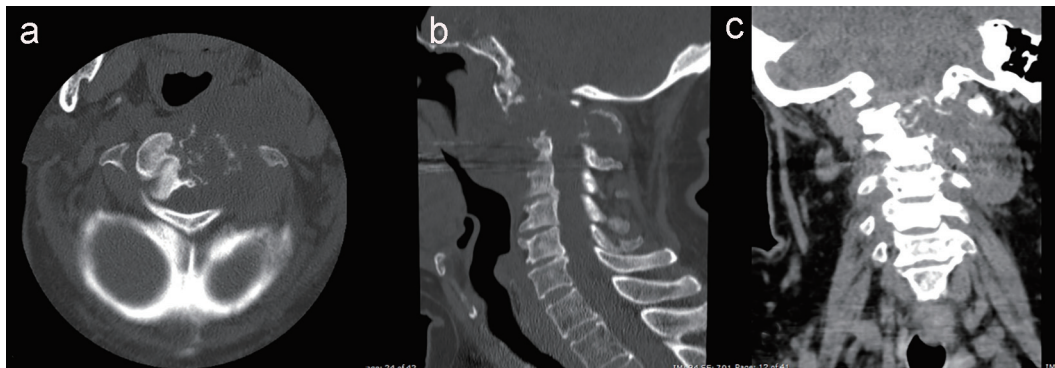


Figure 2. Computed tomography (CT) of expansive and destructive epidural spinal space process, centered in C2 and involving C1 and C3 and infiltrative compromise of neighboring left soft tissues. High enhancement after intravenous contrast injection was observed. (a) Bone window axial view. (b) Bone window sagittal view. (c) Soft tissues window and intravenous contrast, coronal view.

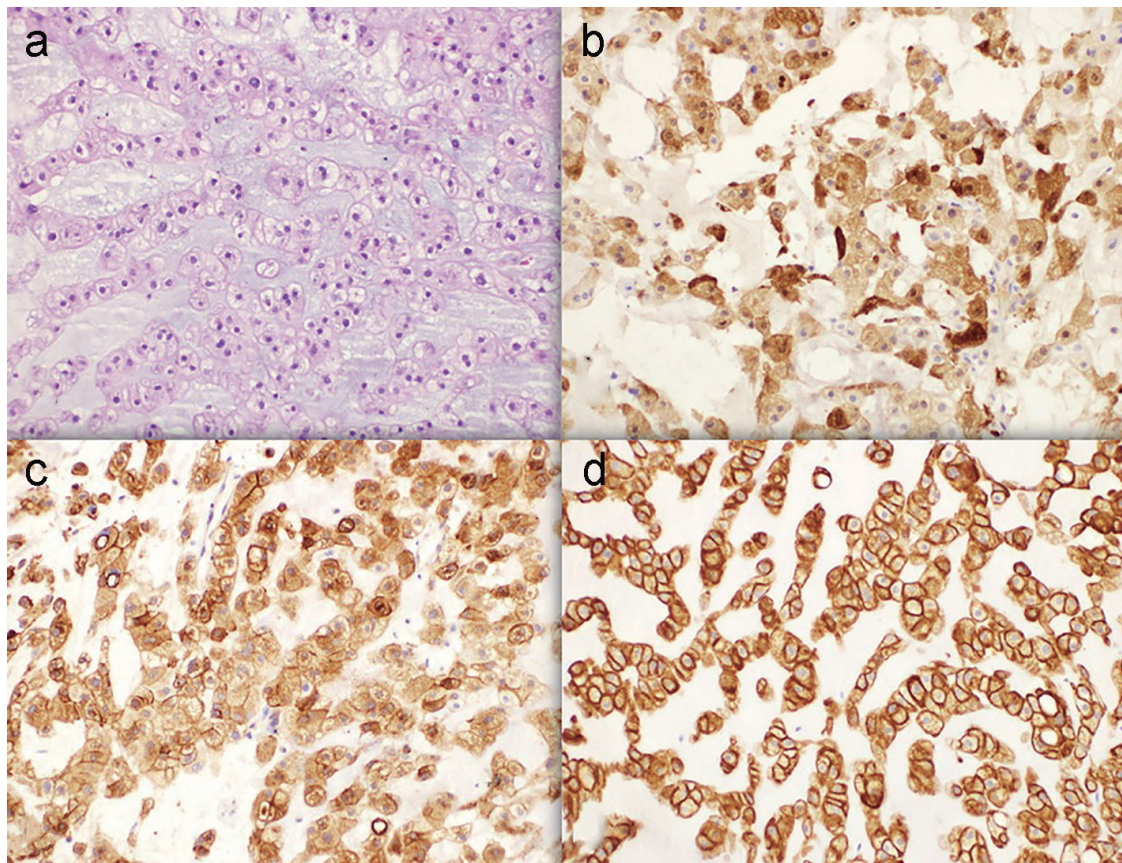


Figure 3. Immunohistochemistry study (× 20). (a) Hematoxylin and eosin stain. (b) Positive S-100 stain. (c) Positive EMA stain. (d) Positive KAE1/3 stain.

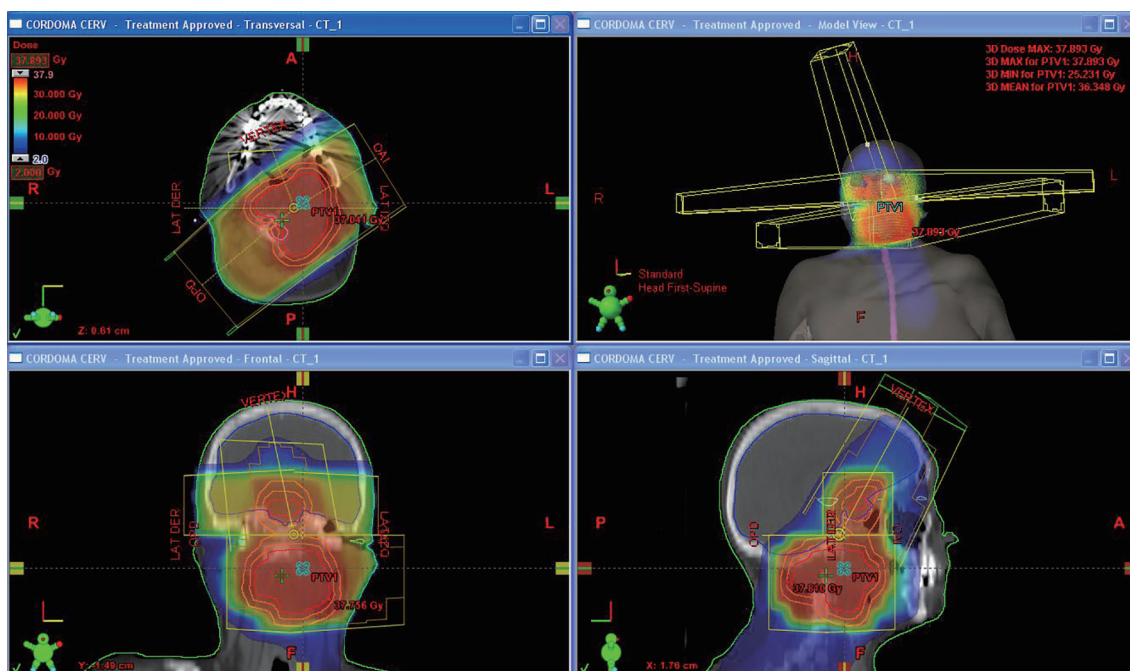


Figure 4. Three-dimensional conformal radiotherapy plan.

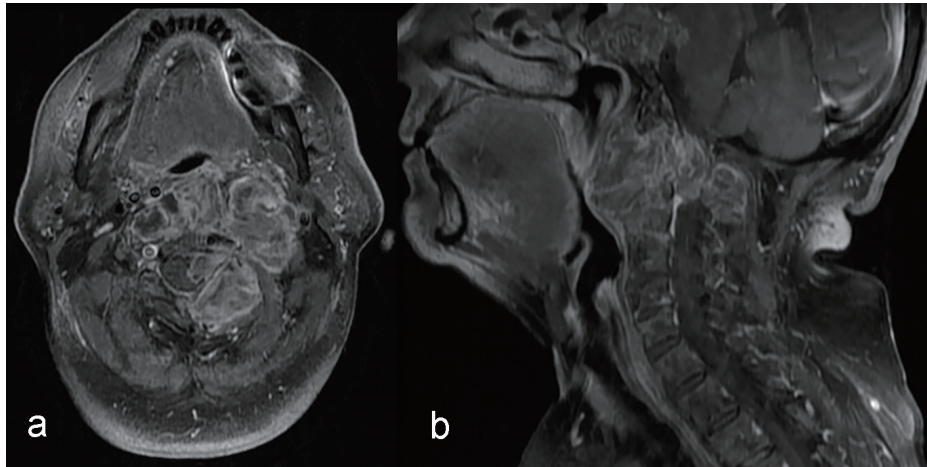


Figure 5. Magnetic resonance imaging (MRI) control imaging after palliative radiotherapy treatment showing growing of the expansive cervical spine process. The mass involved parapharyngeal, retropharyngeal, pharyngeal mucous space and perivertebral spaces, trespassing the middle line and involving the great vessels of the suprahyoid neck. (a) T1 fast sat and gadolinium axial view. (b) T1 fat sat and gadolinium sagittal view.

follow-up is required.

Conclusion

CHCD is a rare entity arising from remnants of embryonic notochord in the craniovertebral axis. Imaging studies play an important role in the treatment planning, providing valuable information about the growing pattern and involvement of critical anatomic organs. The treatment of choice is surgical resection, combined with adjuvant radiation therapy in some cases.

Conflicts of Interest

The authors declare that there are no actual or potential conflicts of interest in relation to this article.

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