Chordomas are rare tumors of notochordal tissue remnants and most frequently affect the sacrum and skull base. We report a case of a 58-year-old woman having cervical chordoma causing related symptoms for 5 years. MRI showed a dumbbell-shape mass with heterogeneous enhancement at C2 and C3 on postcontrast images, mimicking neurogenic tumor. Chordoma should be included in the differential diagnosis of a dumbbell-shaped mass involving upper cervical vertebrae in patients with long duration of symptoms.

Key words: Cervical spine; Chordoma; Spinal tumor

Chordomas are rare and slow-growing destructive tumors that are histologically benign but locally invasive in behavior. The tumors arise from the developmental remnants of the primitive notochord. The incidence of the tumors peaks in age of 50 to 60 years with male predominance (male: female = 2:1) [1]. The tumors account for 1 to 4% of all primary malignant bone neoplasms. Approximately 50% of the cases arise in the sacrococcygeal region and 35% arise in the skull base. The remaining 15% of tumors occur on the midline along the path of the notochord, primarily involving the cervical vertebrae [2]. Symptoms occur varying with location. The treatment of choice is a wide en bloc excision.

We herein report a case of a 58-year-old female with a histological proved chordoma involving cervical vertebrae.

CASE REPORT

A 58-year-old female complained of progressive neck pain, limb weakness and general numbness for 5 years. She had a history of rectal adenocarcinoma, Duke’s B, and had received lower anterior resection. The serum carcinoembryonic antigen (CEA) levels revealed normal. Physical examinations showed a grade-5 muscle power of four limbs and decreased sensation of pain and temperature of trunk, upper and lower extremities. Increased deep tendon reflex with bilateral positive Babinski signs was noted. Laboratory tests and tumor markers were within normal limits.

Plain radiographs of C-spine showed osteolytic lesion at posterior element of C2 (Fig. 1a). MRI examination (1.5-T, Horizon LX, G.E. Medical systems, Wis.) of C-spine revealed a lobulated soft-tissue mass, measuring 4.5 x 4.5 x 3.5 cm, at C2 and C3 levels. The tumor depicted isointense signal on T1WI (TR/TE/excitations, 450/10/2) (Fig. 1b), high signal intensity on T2WI (TR/TE/excitations, 3300/112/2) (Fig. 1c) and strong enhancement on fat-saturation postcontrast T1WI (TR/TE/excitations, 550/10/2) (Fig.
The tumor mass had a dumbbell shape and involved the right neural foramen of C2-3 and right prevertebral space. Obvious cord compression at C2-3 level was noted.

The patient received total excision of the tumor via anterior and posterior approaches by a neurosurgeon. On gross inspection, the tumor was a well-defined, brownish and elastic mass. The cut surface revealed myxoid change and focal hemorrhage. Microscopically, there were physaliphorous (“bubble like”) cells arranged in cords and nests in the mucinous background (Fig. 1f). Histological diagnosis was chordoma. The patient’s symptoms were improved after the operation.

**DISCUSSION**

Chordoma is extradural in nature and is generally associated with extradural extension and bone destruction. Wippold et al. [3] reported 10 patients having cervical chordoma that these patients usually developed pain and weakness in the neck and shoulder, with duration from acute attack to 12-month. The tumor frequently involved C2 to C5 with 90% located in epidurae. Eighty of them had bony erosion or remodeling, 70% extended to perivertebral space, 40% involved intervertebral disc, and 70% had enlarged lesions destroying the neural foramina. Four lesions of their cases also crept along the cervical nerve roots and even enlarged the neural foramina.
mimicking primary nerve sheath tumors on images [3]. Similarly, MRI of our case demonstrated a dumbbell-shaped tumor involving epidurae of right neural foramen of C2-3 and right prevertebral space, resembling a neurogenic tumor.

On MR images, typical chordomas show low to intermediate signal intensity on T1WI and very high signal intensity on T2WI. Administration of gadolinium results in moderate to marked heterogeneous enhancement [3,4]. Areas of high signal intensity on T1WI have been described occasionally in intracranial or spheno-occipital chordomas, and rarely in spinal chordomas. This feature is likely due to high protein content of the myxoid material [5]. Chordomas are usually high vascularity and frequently develop focal calcifications. In our case, the tumor depicted isointense signal on T1WI, high signal intensity on T2WI, and marked heterogeneous enhancement after gadolinium administration. The inhomogeneous intensity pattern can be related to myxoid change and focal hemorrhage, as proved on histology.

Our patient had a history of Duke’s B rectal cancer and, therefore, metastasis in vertebrae should be excluded before surgery. However, the CEA levels were not elevated and no known systemic metastasis was noted. Metastasis in vertebrae usually involves thoracic and lumbar spine and commonly invades pedicles and posterior elements as well. Our patient only had lesions in the cervical spine with related symptoms persisted for more than 5 years. These findings are not considered relating to previous history of rectal cancer.

Metastasis of chordoma was reported to be as high as 25%. The common sites of distant metastases are the lungs, liver, and bone [6]. In our patient, no evidence of distant metastasis was noted at time of diagnosis and surgery.

In summary, we report a patient with a cervical chordoma. Although it is not specific, chordoma should be included in the differential diagnosis of a dumbbell-shaped mass involving upper cervical vertebrae in patients with long duration of symptoms.

**REFERENCES**

頸椎脊索瘤的磁振造影發現：病例報告

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脊索瘤是來自脊索遺跡的罕見腫瘤，主要侵犯頸椎及顱底。我們報告一例58歲女性患有頸椎脊索瘤及相關症狀五年。磁振造影檢查顯示第二和第三頸椎有一個啞鈴狀的腫塊，注射顯影劑後呈現不均勻的影像增強，非常類似神經性腫瘤。若病人在上頸椎呈現啞鈴狀腫塊及患有長期症狀時，脊索瘤應列入鑑別診斷中。

關鍵詞：頸椎，脊索瘤，脊椎腫瘤