Chordoma of the Thoracic Spine
—Case Report—

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Abstract
A 44-year-old woman presented with a thoracic chordoma with intrathoracic extension manifesting as complaints of lower extremity weakness, hypesthesia below the levels of T5–6, and sphincter incontinence. Almost total resection combined with anterior interbody fusion and stabilization was possible through a left transpleural transthoracic approach. She suffered recurrence after 2 years and was considered inoperable. Biopsy revealed a malignant chordoma with no sarcomatous differentiation. Chordoma is an uncommon malignant bone tumor originating from remnants of the embryonal notochord, occurring mostly along the axial skeleton, at the extremity of the vertebral spine, and is least common in the thoracic region. Differential diagnosis is problematic and biopsy is helpful particularly if considered inoperable. Thoracic chordomas of the malignant type manifest as cord or root compression. Classical malignant chordomas must be distinguished from chondroid, benign, or other types of chordomas, since the biological behavior and clinical features are distinct. However, the differential diagnosis cannot be based on histological examination, but long-term follow up is required. Most patients have extradural and intraspinal tissue extension at the time of diagnosis, which makes complete resection impossible. Aggressive surgery without violation of surgical borders is the best choice in the treatment of thoracic chordoma. Thoracic chordoma is a recurring neoplasm and is prone to dissemination and sarcomatous differentiation despite its slow-growing nature.

Key words: chordoma, notochord, physaliphorous cell, thoracic spine

Introduction
Chordoma is a slow-growing malignant tumor originating from notochordal remnants, which accounts for 1–4% of all primary bone malignant tumors, of which 50% occur in sacrococcygeal, 35% in sphenoccipital, and 15% in vertebral locations,12,26,29,31,32,34 the latter being mostly in the cervical region.12,18,19,26,31,32,34 Thoracic involvement is 2–5%.29,31,32,34 Extraosseous chordoma may occur within the sella, paranasal sinuses, larynx, maxilla, and nasopharynx7 or scapula,18 as well as ectopic intradural foci in the pons and spinal canal.7,27,33,36

Chordoma is the only embryonic neoplasm that presents in the later decades of life, most typically in the fifth through seventh decades.2,3,11,17 However, almost all cases in the sphenoccipital and cervical regions occur in the first two decades of life.9,12,19,34 Vertebral chordomas tend to occur in somewhat younger populations and frequently are more aggressive.18,26,31 Chordomas affect men more than women at a ratio of 2.7/1 to 1.6/12,3,5,11,18,29,31,34 Female patients are younger, and have longer duration of symptoms, larger tumors, and shorter mean survival.5

Chordoma may recur sometimes with malignant transformation despite the slow-growing nature, and recurrence commonly involves contiguous anatomic sites, which incapacitates the patient and makes total resection almost impossible.

Case Report
A 44-year-old woman was referred to us from the Department of Neurology on October 27, 1998 with complaints of backache, lower extremity weakness persisting for 6 months, and sphincter incontinence over the last 2 months. Neurological examination found hypesthesia below the T5–6 levels, 3/5 motor function in the lower limbs, bilateral extensor plan-
Fig. 1 Computed tomography scan with contrast medium showing vertebral bone involvement (irregular contours) and an associated left paravertebral mass with mild contrast enhancement, and intraspinal extension of the mass.

Fig. 2 Preoperative sagittal magnetic resonance images revealing intrathoracic and intraspinal extension of the tumor involving T-4 vertebra and abutting aorta appearing as hypointense on the T1-weighted image (A) and hyperintense on the T2-weighted image (B).

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Clonus, and increased deep tendon reflexes. Thoracic computed tomography (CT) demonstrated a hyperdense, destroyed but non-collapsed vertebral body contiguous with a left paraspinous soft tissue mass (Fig. 1). Magnetic resonance (MR) imaging showed a thoracic mass destroying the T-4 vertebral body, which appeared as hypointense on the T1-weighted image and hyperintense on the T2-weighted image (Fig. 2). The patient was initially considered as inoperable. CT-guided fine needle aspiration biopsy performed to confirm the diagnosis proved to be a malignant tumor.

The patient underwent surgery using a left transpleural transthoracic approach on November 5, 1998. T-4 corporectomy, tumor resection, and spinal cord decompression were performed followed by interbody fusion using a bone graft with a piece of rib and long cervical plate application. Postoperatively hemothorax complication developed, so reoperation was performed on November 14, 1998. The aorta had been lacerated by one of the screws slightly coming out from the plate, and was secured with a Teflon felt barrier. Postoperatively sphincter incontinence and limb weakness improved. The patient did well postoperatively and was discharged on December 2, 1998. Postoperative follow-up MR imaging was performed periodically (Fig. 3). A large recurrent tumor at T-4 level contiguous with a right paraspinous mass (opposite site) was discovered at the 2-year follow up (Fig. 4). The mass was evaluated as inoperable. A biopsy was taken to rule out sarcomatous metaplasia. Radiotherapy was planned for the last step.

Macroscopically, the extrinsic parts of the tumor outside the bone appeared yellow and fatty, and consisted of encapsulated small nodules separated by thin septa. The involved bone was yellow with a cheesy consistency. Histological examination found large epithelioid cells arranged in a cord-like fashion in a mucinous stroma with occasional cells containing mucinous material. The lobulated appearance along with cording of the cells and occasional physaliphorous (large vacuolated) cells suggested the diagnosis of chordoma. Vimentin, S-100 protein, neuron-specific enolase (NSE), cytokeratin, and epithelial membrane antigen (EMA) staining were positive confirming the diagnosis of chordoma (Fig. 5).
Thoracic vertebral chordomas are distinctly uncommon. Utne and Pugh\textsuperscript{32} (1955) had 2/72 cases among all the total chordomas, Higinbotham et al.\textsuperscript{18} (1967) 2/46 cases, Cummings et al.\textsuperscript{11} (1983) 0/24 case, Volpe and Mazabraud\textsuperscript{34} (1983) 0/25 case, Rich et al.\textsuperscript{29} (1985) 1/48 case, O’Neill et al.\textsuperscript{26} (1985) 0/34 case, Amendola et al.\textsuperscript{2} (1986) 1/21 case, Azzarelli et al.\textsuperscript{3} (1988) 0/33 case, and Coffin et al.\textsuperscript{9} (1993) 2/12 cases; whereas Sundaresan et al.\textsuperscript{31} (1979) had 2/54 cases among all the spinal chordomas, Meyer et al.\textsuperscript{23} (1984) 3/21 cases, Firooznia et al.\textsuperscript{13} (1986) 0/16 case, and Bjornsson et al.\textsuperscript{5} (1993) 7/40 cases.

Luschka in 1856 described jelly-like growths surrounding the clivus, and Wirchow\textsuperscript{35} provided a histological description in 1857 as “ecchordosis physaliphora.” Finally Congdon\textsuperscript{10} described “benign chordomas” as of the same origin, i.e. notochordal nests. Benign chordomas are lobulated.
neoplasms consisting of large mucus-containing physaliphorous (large vacuolated) cells and their precursors (elongated epithelioid); whereas other types of chordomas have atypical, epithelioid, or spindle cell or chondroid features. The light and electron microscopy appearance and immunostaining and ultrastructural findings are entirely similar, but classical malignant chordomas, like our case, must be distinguished from other types of chordomas since they are considered to have different biological behaviors. Long-term clinical follow up is usually needed to identify the nature of the chordoma.

Chordoma is composed of mostly epithelial, with less well-developed mesothelial features. Positive staining of epithelial components (physaliphorous cells) for cytokeratin (Cam 5.2) and EMA are helpful in the differentiation from chondrosarcoma. Positive staining of mesenchymal elements for S-100 protein, vimentin, NSE, and fibronectin differentiate chordoma from mucus-producing carcinomas. Positive 5-nucleotidase staining can exclude chondroid chordoma, chondrosarcoma, ependymoma, and ecchordosis physaliphora. Sarcomatous metaplasia occurs frequently in recurrent and radiated lesions. Our patient did not receive radiation therapy, and suffered recurrence but without metaplasia. Carcinoembryonic antigen, glial fibrillary acidic protein, Leu-7, and MAK-6 negativity should be demonstrated when needed.

Chordomas may occur without bone and disc involvement. However, malignant chordomas are usually extradural and cause local bone destruction. Chordomas are well demarcated from the soft tissue due to the fibrous pseudocapsule, but have elusive margins in bone. Radiography shows osteolytic lesions in the vertebral body, extension into the adjacent vertebra destroying the intervening disk space, secondary reactive osteosclerosis (not osteoblastic activity but vertebral collapse), and characteristic amorphous soft tissue calcifications or ossifications. CT clearly demonstrates ivory vertebral body and paravertebral tumor. Contrast enhancement is mild to moderate and heterogeneous (mottled). Septated areas of low attenuation within the tumor or multiple zones of hypodensity-correlating areas of cystic degeneration may be present. MR imaging shows epidural extension, compression, and epidural metastasis. The tumor is hypointense or isointense to the neural tissue on T1-weighted images, and may include areas of hyperintensity due to mucin or hemorrhage; and hyperintense on T2-weighted images. Contrast enhancement is heterogeneous and moderate to marked. Angiography and preoperative embolization may be helpful in some cases.

The differential diagnosis between typical and chordoid chordomas cannot be established by radiological methods. In the presence of destructive changes of multiple adjacent vertebrae and well-defined soft tissue mass, as in our case, the differential diagnosis includes chondrosarcoma, metastasis, lymphoma, myeloma, and occasionally granulomatous osteomyelitis. The tumor in our patient was presumed inoperable. Later biopsy proved malignant chordoma.

Unlike benign chordomas (if not intraspinal), chordoid chordomas, and malignant chordomas...
manifest as different symptoms according to the location.\textsuperscript{18,28–31} Thoracic and lumbar chordomas, as in our case, manifest as symptoms of cord or nerve root compression.\textsuperscript{29–31} Paraplegia or quadriplegia occur only late in the course.\textsuperscript{30,31}

The more accepted approach is aggressive surgery\textsuperscript{25} to resect the tumor as far as possible and prevent dissemination into the surrounding tissues.\textsuperscript{15} Thoracic and lumbar chordomas are more difficult to resect totally than sacral lesions, since most patients like our case have extradural and paraspinal tissue extension at the time of diagnosis.\textsuperscript{3,17,31} Most previous posterior debulking approaches have been replaced by anterior approaches to assure complete resection.\textsuperscript{21} Our patient received no radiotherapy and is considered to be directly related to the surgery\textsuperscript{25} to resect the tumor as far as possible and preclude violation of the tumor margins at the time of resection.\textsuperscript{21} We performed the same intervention, which was quite successful.

Recurrence after surgery is common (28–68\%), and is considered to be directly related to the surgical violation of the tumor margins at the time of resection.\textsuperscript{21} Our patient received no radiotherapy and suffered recurrence after 2 years. Despite radioresistance, palliative low-dose or hyperfractionated high-dose radiotherapy,\textsuperscript{2,3,17,18,23,31} or ion beam or interstitial therapy\textsuperscript{2–17} is recommended following total resection or palliative debulking.\textsuperscript{8,24,31} Radiotherapy does not prevent recurrence, or provide tumor regression,\textsuperscript{2,3,31} but may increase the disease-free survival. Since chemotherapy is considered to be ineffective,\textsuperscript{3,26,31} or needs further investigations,\textsuperscript{26} we did not attempt this therapeutic modality.

Metastasis is encountered in 3–60\% of cases.\textsuperscript{5,8,18,19,24,26,31,37} Chordomas of the mobile spine metastasize more often than sacrococcygeal lesions,\textsuperscript{10} to the skin, lung, lymph node, bone, liver, and other intraabdominal viscera.\textsuperscript{3,8,9,16,21,25,31,37} Intradural metastasis\textsuperscript{21} and subarachnoid dissemination\textsuperscript{1} may also occur. Our patient was metastasis-free at the time of second admission. Complete cure is rare; survival is relatively short for patients with tumors in a vertebral location. Five-year survival is reported to be 10–70\% of patients with chordoma.\textsuperscript{5,8,17,26,29,31}

References

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