Chordoma of the thoracic spine in an 89-year-old

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Abstract

Study design Case report and literature review.

Objective Report the highly unusual presentation of thoracic spine chordoma in an 89-year-old and review existing literature as it impacts treatment in the elderly.

Summary of background data Chordomas are infrequent tumors of the spine that commonly present during mid-adulthood at the spheno-occipital or sacrococcygeal junctions. The mobile spine is affected in 10–15% of cases but chordomas are extremely rare in the thoracic spine. Chordoma rarely enters the differential diagnosis of spinal tumors in elderly patients, for whom metastases and multiple myeloma are by far the most common.

Methods A case report is detailed of an 89-year-old male presenting with incapacitating pain and early signs of thoracic myelopathy. A lytic, expanding lesion of the T10 vertebral body with epidural spinal cord compression was identified. In the absence of evidence of other primary tumor, a CT-guided needle biopsy revealed chordoma. A literature review of reported thoracic spine chordomas was also performed.

Results An intralesional posterolateral resection and reconstruction was performed with good results and no recurrence at 13 months' follow-up. 30 reports of thoracic spine chordomas were identified in the literature since 1902. Mean age of presentation at 35.7 years is earlier than for most chordomas. The oldest previously reported patient was 68-year-old. Neurological impairment at presentation is rare with the usual presenting symptom being pain.

Response to radiation and chemotherapy is limited. 5-year survival rates range from 50 to 60%.

Conclusion Chordoma is a highly uncommon epidural neoplasm of the thoracic spine and is vanishingly rare in elderly patients. This report documents the oldest reported patient with thoracic spine chordoma at 89-year-old. Their locally aggressive behavior typically prompts consideration of aggressive surgical resection. When tailored to the individual patient, such procedures are feasible without excessive morbidity even in elderly patients.

Keywords Chordoma · Spine · Vertebrae, thoracic · Corpectomy

Introduction

Chordoma is an unusual disease accounting for only 1–5% of all osseous neoplasms [1, 2] but nevertheless remains the most common primary neoplasm of the spinal column. Prompt recognition and treatment of this condition is warranted since its clinical course and treatment are markedly distinct from the much more common diagnosis in the differential for spinal column tumors, namely, metastases from other primary malignancy [3].

Chordomas are thought to arise from notochordal remnants due to their histological similarity but extra-spinal foci, such as the larynx, maxilla and skin have also been described [2, 4]. Despite the putative embryological derivation, the most commonly affected age group is middle-aged adults with most series listing the average age at presentation in the sixth decade [2, 3, 5, 6]. Chordomas arise preferentially at two distinct sites along the spinal axis: the sacrococcygeal (50–60% of all cases) and sphenooccipital (35–40%) junctions [1, 7]. Chordomas arising

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from the so-called “mobile” spine are much less common and account for less than 15% in most series [3]. Particularly rare among these mobile spine chordomas are those originated in the thoracic region.

The objective of this case report is to both describe an extraordinarily rare presentation of chordoma in the thoracic spine in a patient at the end of the ninth decade of life and to review the existing literature on thoracic spine chordomas as well as therapeutic options for elderly patients with such an uncommon diagnosis.

Case report

An 89-year-old male presented with progressively severe mid-thoracic back pain of 6 months duration and more recent decline in ambulatory function and bladder control. Past medical history was positive for coronary artery disease and cardiac dysrhythmia, for which he had two coronary stents and a defibrillator (AICD) implanted. Initial workup at an outside institution included thoracic spine computed tomography (CT) and a needle biopsy, after which he was transferred to our hospital. At that time, neurological examination revealed severe gait imbalance and lower extremity hyperreflexia. Cognition, motor, and sensory exams were all normal. He had been completely independent until 2 months prior to admission, after which he required increasing assistance due to his worsening ambulatory status, correlating with a decline in his Eastern Cooperative Oncology Group (ECOG) performance status score from 0 to 3. Magnetic resonance imaging was contraindicated because of the AICD, therefore, a CT-myelogram with sagittal and coronal reformatted images was performed instead. This study demonstrated a highly lytic lesion involving the T10 vertebral body with epidural extension into the spinal canal with some rostral and caudal extension within the canal. Circumferential disease was already present at that time, both in the posterior elements of the vertebral segment and in the epidural space. Spinal canal diameter was also severely diminished producing significant spinal cord compression with near myelographic block (Fig. 1). Diffuse idiopathic skeletal hyperostosis (DISH) was also evident. The material obtained from the original CT-guided needle biopsy of the T10 vertebral body was re-examined at our institution. In spite of the atypical location, histology and immunohistochemistry were consistent with chordoma and independently agreed upon by pathologists at both the referring hospital and our institution [8]. Due to the patient's advanced age and medical comorbidity, the radiation oncologists (including those from a proton beam center in a neighboring state) were initially consulted. They, however, felt that radiation therapy alone was unlikely to provide substantial benefit

Fig. 1 **a** (axial), **b** (sagittal) Preoperative CT–myelogram showing T10 lytic lesion, with epidural extension into the spinal canal and marked cord compression. Note anterior bridging osteophytes that confer additional stability to segment. **c** (axial), **d** (sagittal) Postoperative CT scan at 4 months visit showing satisfactory decompression and tumor resection. Alignment is maintained and the vertebral body defect was adequately filled by cement

given the size of the tumor and its proximity to the spinal cord as well as the variable response of chordoma to even proton-based treatments.

Faced with an uncommon diagnosis in a highly functioning elderly patient, a decision was made to proceed with surgical treatment. Cortical wall violation and epidural spread of the tumor essentially negated the option of true en bloc spondylectomy. Therefore, the patient underwent a posterolateral intralesional resection via a bilateral transpedicular approach with near total corpectomy. Only the anterior and lateral cortical walls of the vertebral body appeared free of tumor. A purple-tan, moderately vascular neoplastic mass encircled the dura. All gross tumor tissue was successfully removed piecemeal. For reconstruction, methylmethacrylate bone cement was carefully applied to the corpectomy defect extending from the endplates of the adjacent vertebrae and allowed to harden. Based on the preoperative finding of DISH, autofusion was assumed and was confirmed at the time of surgery by manipulating the spinal elements from T9 to T11. Therefore, no instrumentation was inserted. Pathology confirmed the diagnosis

of chordoma with an immunohistochemistry profile positive for cytokeratin 8/18 and epithelial membrane antigen (EMA) while negative for S-100 protein (identical to the previous biopsy) [8]. The patient recovered uneventfully, and he was transferred to a rehabilitation facility on the fifth postoperative day. He subsequently underwent photon-based intensity-modulated radiation therapy (IMRT) due to logistical constraints associated with interstate travel for proton beam therapy. He received a total dose of 5,040 cGy over 28 fractions of 180 cGy daily with the use of image-guided radiation therapy. He suffered no radiation side effects. After 13 months of follow-up thus far, the patient has demonstrated full recovery of ambulation, normal control of micturition, and ability to independently perform all activities of daily living, thus improving his ECOG performance score to 1. Postoperative CT at this early time point shows neither evidence of tumor recurrence or change in spinal alignment (Fig. 1c, d).

Discussion

Thoracic spine chordomas are a rare entity. An extensive review of the English-language literature from 1902 onward yielded only 17 reports totaling 30 cases (Table 1) [1–4, 6, 7, 9–19]. The last literature review on the subject

identified only eight cases [11]. These tumors represent only 8–17% of mobile spine chordomas and an estimated 1% of all chordomas [2, 3]. A peculiar characteristic of this patient group is that thoracic chordomas tend to arise at a much earlier age than expected with mean age at presentation only 35.7 years (Table 1). The previously reported oldest patient was 68 years old [13]. A distinct peak at an unusually young age for chordomas can also be identified: three patients were 15 years or younger at presentation. Gender prevalence is similar to chordoma in general with higher rates in males (13 male:7 female in thoracic spine compared to 2 male:1 female in general chordoma). Another interesting characteristic of the thoracic chordoma subgroup is that only 2 of 30 reported patients presented with neurological deficits. The most common complaints at presentation were chest or back pain, followed by anterior mediastinal compression syndromes (hoarseness, dysphagia, cough). Up to a third of the reported patients were entirely asymptomatic and diagnosed incidentally on routine chest radiographs. The patient presented here similarly started predominantly with painful complaints that only just prior to presentation began to produce neurological symptoms despite advanced vertebral body destruction and epidural compression of the spinal cord.

Because of the small number of cases reported and the highly variable treatments employed since the first report in

Table 1 Literature summary

Ref #	Period/year reported	# pts mobile spine	# pts thoracic spine (%)	Gender	Age	Level	Symptom
1	1949–1976	18	2 (11%)	–	–	T?	–
2	1954–2002	52	4 (7.7%)	–	–	T4, T9, T10, T11	–
3	1902–1993	40	6 (15%)	6 M, 1 F	–	T?	–
4	2002	1	1	F	44	T4	Paraparesis
6	1963–1998	12	1 (8.3%)	M	40	T1 + T12	–
7	2003	10	1 (10%)	–	–	T7	–
8	1992	1	1	M	9	T3	XR finding
9	1956	2	2	2 M	37	T4	Chest pain
					43	T4	Back pain
10	1983	1	1	F	14	T4	XR finding
11	1953–1987	14	4 (29%)	–	–	T?	–
12	2008	1	1	F	68	T7	Upper abdominal pain
13	1966	1	1	M	46	T2	Cough
14	1998	1	1	F	39	T2	Hoarseness
15	1985	1	1	M	59	T8	Paraparesis
16	1996	1	1	F	15	T3	XR finding
17	2008	1	1	F	25	T2	XR finding
18	1951	1	1	M	30	T4	Dysphagia

Previous published literature review yielded only eight cases

ref # reference number, # pts number of patients, – data is not cited, T? unspecified thoracic level

1902, it is difficult to accurately describe the natural history and treatment responses of thoracic spine chordomas. Both radiation- and surgery-based treatment modalities have evolved considerably during this time. Furthermore, not all reports include patient outcomes. Nevertheless, it appears that despite the earlier age of symptom onset, thoracic spine chordomas are not more aggressive than other chordomas. Data from the two largest series indicate survival rates between 50 and 60% at 5-year follow-up after surgical resection and radiation therapy [2, 3, 9]. Metastatic disease occurs in 5–10% of all chordomas but is rarely present at the time of diagnosis [2, 6].

Chordomas are known to be locally aggressive and relatively unresponsive to chemotherapy and radiation [20]. Aggressive resection followed by proton beam therapy has evolved as a popular treatment strategy, and whenever possible, en bloc resection techniques yield the longest disease-free and overall survival [2, 3]. En bloc spondylectomy and even intralesional resection however are not devoid of complications. Of the 15 reported cases that detail outcomes, 2 died from perioperative complications [13, 15]. As always, the balance between treatment morbidity and long-term disease control must be carefully discussed with the patient. Fortunately, advances in surgical techniques and perioperative care have made aggressive resections safer. In that regard, our case not only illustrates the oldest reported patient with a thoracic chordoma but also an example of a successful aggressive treatment strategy in a patient who may not have been considered a surgical candidate due to age and comorbidities. Posterolateral approaches (e.g., transpedicular or costotransversectomy) for resection of vertebral body tumors have evolved as safe and effective means of spinal cord decompression while enabling posterior reconstruction/stabilization with the same exposure [21, 22]. These are ideally suited to the thoracic region of the spine where ligation of a nerve root can be accomplished with minimal morbidity to access the ventral spinal elements and where avoidance of anterior thoracotomy reduces perioperative morbidity substantially. Admittedly, corpectomy with intralesional resection may confer only moderate improvements in survival compared to palliative laminectomy or radiation therapy alone, but as in the case presented here, definitive en bloc spondylectomy is not anatomically feasible in all patients. In this particular case, cement reconstruction of the anterior column provided immediate support with low cost and ease of application while this patient's particular anatomy obviated the need for instrumentation. We feel that the rationally designed treatment approach described here has conferred substantial benefit for maintenance of quality of life and function (as reflected in improved ECOG score) while still being mindful of the overall life expectancy of our unique patient.

Conclusion

Thoracic spine chordomas are truly rare neoplasms, particularly compared to metastatic lesions of the spine. As reflected in the case presented here, however, chordoma should still appear in the differential diagnosis for solitary spinal column tumors even in elderly patients. Since the treatment options for various malignancies affecting the spine range from simple radiation to radical surgical removal, diagnostic biopsy should be attempted on solitary lesions to guide treatment planning. Mortality from chordomas typically results from locally aggressive progression of disease. Therefore, surgery is the mainstay of treatment in patients with good performance status prior to symptom onset and should be strongly considered in patients of any age.

Conflict of interest None of the authors has any potential conflict of interest.

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