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Case Series

Sacral Chordomas; Problems in a Developing Nation with Expensive Healthcare Needs. A Case Series and Discussion of Socioeconomic Problems of Treating Long-term Malignancies in Developing Countries

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Abstract

Chordomas of the mobile spine (C3 to L5) constitute less than 5% of the overall incidence of chordomas in the spine. They generally are osteodestrictve leading to vertebral collapse and severe deficits including paraplegia and quadriplegia. We present a case series of 3 sacral chordomas operated in a period of 1 year from 2017 to 2018 in a tertiary care center in southern India. Apart from clinical presentation and surgical clearance, we present our take on challenges faced by patients of extensive spinal malignancy to finance and care for their disease in a developing country bereft of state assistance in healthcare.

Keywords: Chordoma; Mobile Spine; Tuberculosis; Canal Stenosis; Instability; Laminectomy

Introduction

Spinal chordomas are rare, constituting only 10 - 15% of the incidence of chordomas in toto. The lesions are seen most frequently in the sacrum (80%) followed by the Craniocervical junction (15%) chordomas across the rest of the mobile spine are extremely rare and present as destructive lesions leading to collapse of the affected vertebral body leading to instability and pain [1,2]. These lesions rarely cause radiculopathy and canal stenosis by themselves without bone destruction. Intradural extensions are almost unheard of [2].

We present a case series of 3 sacral chordomas operated in a period of 1 year from 2017 to 2018 in a tertiary care center in southern India. Apart from clinical presentation and surgical clearance, we present our take on challenges faced by patients of extensive spinal malignancy to finance and care for their disease in a developing country bereft of state assistance in healthcare.

Case Series

Case 1

A 54-year-old lady presented with complaints of low back ache radiating to both lower limb (right much more than left) for 3 months. She also complained of Weakness in both lower limbs for 2 months, coupled with loss of urinary continence for 2 months. The patient was apparently normal 3 months ago when she developed low back ache which was insidious in onset, gradually progressive and seen radiating to both lower limbs. The pain was aggravated on walking and relieved on taking rest and was associated with tingling sensation along the whole length of the lower limb. The patient also had weakness of both lower limbs which manifested mainly as difficulty in getting up from sleeping or sitting position and the need of support for walking. Distal muscle weakness manifesting as a history of slippage of footwear with her knowledge was present as well. For the past 2 months, she complained of urinary

incontinence with increased frequency and urgency. She does not have any comorbidities such as diabetes, hypertension, chronic kidney disease, heart disease or tuberculosis. On neurological examination, her Upper limbs-had normal power. Her Lower limbs had power-Right 4/5; Left 4+/5 with right EHL weakness coupled with restriction of both SLRT at 45°. Both ankle jerks were decreased to 1+ and plantars were mute. Sensations were normal except for a loss of perineal sensation. No spinal deformity, local tenderness or paraspinal muscle spasm were present.

MRI of the lumbosacral spine showed a Large ill-defined mixed intensity lesion causing significant destruction of sacral bone on right side extending from S2 and involving all sacral vertebral bodies inferiorly with minimal post-contrast enhancement. A CT scan of the lumbosacral spine showed an ill-defined lytic lesion in the sacrum involving all the sacral vertebrae on the right extending into the midline and partly to the contralateral side. Cortical destruction with extraosseous soft tissue component. The lesion extends from right sacral ala up to coccyx, also involving inferior articular facet of right ilium, and anteriorly involving piriformis muscle, retroperitoneal space and mesorectal fascia infiltration (Figure 1).

Figure 1: (A) showing a plain radiograph of the sacrum in AP direction, demonstrating extensive destruction of the sacrum by the lesion. In (B) an axial CT scan of the sacrum shows the extent of the destruction where bone is replaced by tumor tissue.

Patient underwent sacral tumor excision and spinal stabilization with iliac and L4, L5 pedicle screws and rods construct under GA. Post-op CT Spine showed implants in place. Histopathology (HPE) was suggestive of a Chordoma with Immunohistochemistry (IHC) showing EMA, S-100 and CK positivity (Figure 2). In view of HPE report, the patient was advised radiotherapy. The patient improved symptomatically and was discharged ambulatory, with no fresh neurological deficits, with Foley's catheter in situ in view of bladder involvement.

Figure 2: Histopathology H&E section in low and high magnification shows large epithelioid cells with central bland nuclei and abundant eosinophilic, finely vacuolated cytoplasm.

Case 2

A 46 year old male patient presented with complaints of low back ache for 6 months, radiating to both lower limbs, coupled with urinary incontinence and constipation for 5 months. The patient was apparently normal 6 months back when he started having low back ache which was insidious in onset, gradually progressive, of a dull aching type which radiated to both lower limbs along the posterior medial aspect of the thigh. The pain was aggravated for 1 month with no relieving factors. The patient also complained of urinary incontinence for 5 months. He initially had urge incontinence but now can't feel fullness of bladder or passage of urine. There is associated constipation for 5 months, which has progressed to such an extent that the patient is unable to defecate freely and must evacuate stools with digital assistance. He is a diabetic and hypertensive on poorly supervised treatment for 1 year. No previous major surgeries or drug allergies are recorded. On Neurological Examination, Lower limb power was bilaterally 4+/5 with EHL normal. Bilateral ankle jerks were mute with plantars flexor. Cremasteric reflexes were present on both sides. No sensory loss. No cerebellar or meningeal signs.

The MRI of the lumbosacral spine showed a T1 hypointense, T2 heterogeneously intense lesion in sacral region with S2, S3, S4 bony

destruction with compression on the rectum and bladder anteriorly. A CT of the lumbosacral region showed a well-defined expansile lytic lesion arising from S1-S5 vertebrae measuring 83x93x94mm extending anteriorly into the pre-sacral space and posteriorly into the spinal canal with destruction of the spinous processes suggestive of a sacral chordoma (Figure 3).

Figure 3: (A) Axial CT spine of the sacrum showing a large destructive mass lesion occupying the body of the bone with (B) showing post sacrectomy Xray of the pelvis showing the fixation construct with iliac screws.

This patient underwent excision of sacral tumor with fusion of L5-S1 with iliac screws done under GA. Post-operatively, he improved symptomatically with complete recovery of pain. He developed mild wound discharge which was managed conservatively. His HPE report was suggestive of sacral chordoma, due to which he was referred for radiation therapy. He was discharged with his wound clean. The patient was ambulatory with no fresh neurological deficits. The patient still had urinary incontinence and hence was discharged with foley's catheter. Constipation improved with laxatives.

Case 3

A 24-year-old lady presented with complaints of lower back ache for 2 years, with urinary incontinence for 6 months. The patient was apparently normal 2 years ago, when she started having back pain, which was insidious in onset and gradually progressing, a dull aching type with radiation to both lower limbs along the anterior aspect of thigh and leg. The urinary incontinence began 6 months ago. Initially he had stress incontinence which progressed to complete loss of bladder sensation presently. He is a known case of hypertension on poor control. No weight loss of loss of appetite. Neurological examination revealed, Normal power in both upper limbs and lower limbs, with no diminished reflexes and plantars flexor. There was sensory loss over the L5-S1 dermatomes on both sides, along with decreased perianal sensation. On per rectal examination, anal tone was lax with a rectal mass palpable underneath the mucosa. The mucosa was free.

A large heterogeneous lesion involving the L5 vertebral body, and the sacrum was seen on MRI of the lumbosacral region. The lesion was hyperintense on T2, mixed on T1 with hyper and hypointense signals both co existing in a multiloculated appearance. There was intense contrast enhancement seen. A CT scan showed an expansile mass lesion arising from L5, S1, S2 and S3 vertebral body (Figure 4).

This patient underwent a total tumor excision and spinal stabilization under GA. Post operatively patient improved symptomatically. The HPE was suggestive of malignant chordoma. The relatives however refused further treatment was discharged the patient to pursue adjuvant therapy later despite advice to the contrary.

The patient was thus discharged ambulatory, with no fresh neurological deficits.

Discussion

Figure 4: (A) axial Ct of the sacrum showing a large destructive lesion destroying the bone, and (B) the post op Xray showing the stabilisation construct after radical excision of the lesion with reconstruction.

Chordomas develop from notochord remnants and are slow growing, low grade malignant tumors. They have been known to be locally invasive [3,4].

Treatment of chordomas consists of radical excision, stabilization, followed by adjuvant treatments (radiation and possibly chemotherapy) [4]. Based on well-established principles presented in the Enneking classification, resection should involve obtaining a wide margin or performing en bloc resection. It is generally accepted that the resected margin should be free of tumor. Boriani recommend en bloc excision of all spinal chordomas when feasible [3,4].

If a total tumorectomy is not feasible, palliative debulking followed by radiotherapy is worthwhile [4,5]. Although chordomas are known to be relatively radioresistant, the value of radiation therapy has been stressed in several reports and series. As most studies cover long periods, during which new techniques were developed, and as the tumor is rather rare, there is no consensus in the literature regarding the optimal radiotherapy scheme for cervical chordomas [4,5]. Proton-beam and stereotactic radiotherapy are currently being investigated and recommended as treatment for these lesions However, there is no clear evidence about the efficacy of these modalities at this time [5,6]. Chemotherapy does not play a role in the treatment of chordomas, although the use of imatinib mesylate is being investigated [6].

Chordomas are well-known to recur locally [5-7]. The recurrence rate seems to be related to the incompleteness of resection. Chordomas of the mobile spine metastasize more often than sacrococcygeal lesions. The reported incidence of metastases varies widely, from 3% to 60%. They are discovered between one and 10 years after the initial diagnosis. Sites of metastases include bone, lungs, lymph nodes, soft tissues, intrathecal space and liver [6-8].

Only primary and complete resection of the tumor offers a major advantage as far as prognosis is concerned [7,8]. Clearly the length of follow up in the present study is short, but the extent of tumor resection that could be achieved using these techniques, as well as the limited risk of morbidity encountered and the successful stabilization that was accomplished, has been encouraging.

Financing long-term cancer care

Countries such as India with primitive health infrastructure and poor awareness of disease and pathological processes often struggle to handle the burden of serious illness es such as cancer. India is often considered to have a double burden of disease owing to its incomplete and lopsidedly distributed industrialization, which gives rise to the prevalence of infectious diseases and epidemics coupled with the newer entrants of cardiovascular disease and malignancy.

After the shock of initial suspicion, investigations and counselling, an estimate is often sent to the patients for costs covering hospitalization and surgery. This sum in itself is prodigious and beyond the means of 80% of the population. Despite this a herculean effort is made often at the mercy of unscrupulous money lenders and landsharks. As diseases often do, overstay in the hospital coupled with complications overshoot the budgetary estimates made making the situation untenable for most. Incomplete treatment is often seen, where discharge is sought due to absolute penury. Those who complete the initial phase of treatment manage to remain disease free for a while. Approximately 20-30% of those who complete the initial phase of therapy manage to seek further adjuvant therapy and stay in surveillance. Charitable trusts and foundations often come forward to assist in financial support but are a drop in the ocean of financial turmoil engulfing the poor.

Another source of spurious comfort remains health insurance schemes for the poor, which claim to cover all expenses and afford seamless and cashless experiences to the unsuspecting customers. Patients often find claims rejected with alacrity due to bureaucratic bungling or plain indifference. The schemes backed by the central government of India, ensure ridiculously poor remuneration for the hospitals and doctors which fail to even cover the costs of the procedures and stay involved. This leads to mistrust between healthcare providers and patients breaking down the basic pillar of the doctor-patient relationship. An untoward consequence of this is the increasing number of violent altercations between medical staff and patient relatives when facing adverse patient health situations.

Faced with such a predicament, patients often stay away from hospital, doctors and often attempt to seek care from traditional medical care practitioners. These unregulated and poorly defined specialties often cheat the patients promising unreasonable and unattainable cures with traditional remedies laced with illegal substances designed to provide temporary relief but nothing else.

Hence, providing effective multimodal care for malignancies with proper surveillance remains a challenge like none other, to be

faced by doctors and patients alike; A mountain almost as mighty to surmount as the disease itself.

Conclusion

Spinal chordomas are great mimics, resembling infectious and other neoplastic pathologies. A high index of suspicion assists in a quick diagnosis and early treatment and a longer disease free and symptom free survival. Proper counselling of patients with financial assistance and encouragement is necessary for all round care and surveillance to truly beat complex malignancies such as chordomas in the future.

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