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CASE REPORT

Cutaneous Metastasis from Sacral Chordoma: A Case Report

P. Unnikrishnan[1], Sreejith G. Nair[1], M T. Sugeeth[1], Rony Benson[1], Rari P. Mony[2]

[1]Department of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India
[2]Department of Pathology, Regional Cancer Centre, Trivandrum 695011, India

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**Corresponding author:*
M T. Sugeeth, MD, DNB, DM, MNAMS
Assistant Professor, Department of Medical Oncology,
Regional Cancer Centre, Trivandrum 695011, India.
Tel: +91-9496324153
Email: sugeethmt@ymail.com

ABSTRACT

Chordoma is a malignant, slow growing and locally aggressive tumor. It arises from remnants of the notochord and accounts for 1–4% of all primary bone tumors. They usually arise from anywhere along the spine, from base of the skull to the sacrococcygeal area and usually do not metastasize. Chordomas are slow-growing tumors which are not responsive to conventional chemotherapy or radiation. They are usually diagnosed late in the course of the disease, as they are low-grade tumors that show a slow progression. Complete surgical excision is the only therapeutic modality to offer a cure. We present a 48-year-old man with progressive paraparesis and disseminated cutaneous and lung metastases diagnosed as metastatic chordoma of the sacrum.

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Introduction

Chordomas are aggressive, locally invasive rare tumors which develop from intraosseous embryonic notochord remnants and therefore occur in the midline, vertebral column most commonly in the sacrum as slowly progressive low-grade tumors.[1] The incidence of chordoma increases with age. They are usually diagnosed late in the course of the tumor growth causing widespread bone destruction and invasion to the adjacent neurological structures resulting in neurological deficits.[1] The incidence of metastasis from chordoma ranges from 3 to 48%, observed more commonly with primary localized vertebral chordomas than with sacrococcygeal chordomas.[2] Skin metastasis from chordoma is extremely rare with only very few cases reported so far in the literature.[3,4] Sacral chordomas presenting as skin metastasis is even rarer.[5] We present a man with a history of traumatic paraparesis presenting with worsening neurological deficit and disseminated cutaneous and lung lesions.

Case Report

A 48-year-old man with a history of paraparesis following a road traffic accident at 25 years of age presented with extensive subcutaneous lesions of forearms, left thigh and low backache of 8 month duration. He also had worsening of lower limb force and urinary incontinence for the last six months. There was no history of fever and weight loss. According to "Eastern Cooperative Oncology Group performance status", his score was 4 at presentation.[6] Physical examination showed firm subcutaneous swellings of 7×7 cm in both forearms, 10×6 cm in left thigh and another swelling in lumbosacral region measuring 3×3 cm. In addition to decreased force of lower limbs, sensory loss below L1 dermatome with loss of bowel and bladder function were observed.

MRI of lumbosacral spine showed an expansile lobulated

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lesion measuring 17×21 cm in presacral, posterior paraspinal and left anterior paravertebral region with extension into spinal canal up to L2 level compressing cauda equina nerve roots along with erosion of sacral vertebral bodies, involving iliac bones and iliopsoas muscles. Biopsy from the cutaneous swellings showed nests of physaliferous cells. On immunohistochemistry, the cells were positive for CK and S100 protein. These features (pathology and IHC) were diagnostic of metastasis from chordoma. Further imaging studies including computed tomography scan of the thorax showed also multiple pulmonary metastases. CBC, serum electrolytes, renal and liver function tests were within normal range. Serum LDH was 369 IU/L. In view of the extensive disease and poor performance status, the patient refused to undergo chemotherapy and was planned to receive supportive care.

Discussion

Chordoma is a malignant, slow growing and locally aggressive tumor that arises from remnants of the notochord. It is generally a rare tumor with an incidence of about 0.1/100,000 that accounts for 1–4% of all primary bone tumors. it is considered the fourth most common malignant bone tumor and the most common primary malignant tumor of the sacrum.[1] Chordoma is a locally destructive tumor commonly observed in the sacrum (50–60%), base of the skull (25–30%), cervical (10%) and thoracolumbar vertebrae (5%). The clinical symptomatology is determined by the location of the primary tumor and its expansion to the adjacent anatomic structures.[7] The incidence of metastasis varies from 3 to 48% and the most common sites include pulmonary and lymph nodes followed by liver and bone,[1] while skin metastases are described to be rare, with estimated less than 20 cases reported.Riesco-Martínez et al. reported a 45-year-old woman with skin metastasis from a sacral chondroid chordoma. In their case, although soft tissue metastases at L4, liver, bone, and lung metastases were treated with chemotherapy, the patient developed nontender, erythematous nodule on her left cheek six years after partial sacrectomy for her sacral chordoma; the patient died a year after.[4] Collins and colleagues have also reported an incidentally discovered cutaneous metastasis on the right upper back of a patient with a history of sacral chordoma 12 years earlier.[3] Gleghorn et al. reported a 61-year-old man with a scalp nodule, presented 8 years after local excision of sacral chordoma.[5] D'Amuri et al. also reported a case of cutaneous metastasis on scapular region of a 68-year-old woman with a history of excision and radiotherapy for sacral chondroma 10 years earlier.[8] Although the cases presented above confirm that cutaneous metastasis is a rare presentation in sacral chondroma, the presented cases differ from that of ours in one important aspect. Our case was not previously diagnosed with chordoma and most of the patient's symptoms were attributed to the history of road traffic accident; however, presentation of subcutaneous swellings in both forearms, left thigh, and lumbosacral region in addition to spine imaging and histopathologic findings helped us diagnose skin metastases from sacral chordoma.

Chordoma is surrounded by a connective tissue-like pseudocapsule and infiltration of the capsule by chordoma cells and its growth into the adjacent tissues is often found histologically.[7] The treatment of choice for localized chondroma is radical resection with wide surgical margins, but because of the spinal localization and destructive growth pattern of the tumor, as well as the fact that these tumors are diagnosed at advanced stages, where prognosis is poor, radical surgical resection is often impossible and local relapses may be observed frequently.[1] Radiotherapy and chemotherapy are also used as a complementary intervention, while the disease progression limits the efficacy of these treatments and impairs the prognosis.[1]

Conclusion

Chordoma is a rare tumor mostly originates from sacrum notochord. Metastasis is a common observation; however, skin metastasis is rarely reported. Few cases of cutaneous metastasis from sacral chordoma have been reported in patients with known history of chordoma, while in our case, sacral chordoma was diagnosed after skin metastases were identified histologically.

Conflict of Interest: None declared.

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