Case Report

The effect of chemotherapy on sacral chordoma in low resources country: A case report of a rare disease

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ABSTRACT

Chordoma is a rare malignant tumor that is more prevalent among males than females. The diagnosis is usually confirmed by imaging and histopathological examination. Surgery is the mainstay of therapy but in some conditions such as metastasis chordoma or poor patient's health, radiotherapy and chemotherapy can be carried out. We reported a case of metastatic sacral chordoma in a 58-year-old male patient, in which chemotherapy was chosen. Chemotherapy was given three times but did not show significant results.

Key words: Chemotherapy, Chordoma, Malignancy, Sacral chordoma

Chordoma is a malignant, slow-growing, locally aggressive tumor occurring most commonly in the sacrum (50%) followed by the cranium (30%) and the spine (20%). These tumors arise from the embryonic notochordal remnants, along with the axial skeleton. Of all bone malignancy tumors, it has a prevalence of 1–4%. Chordoma is dominant in the Caucasian population where the prevalence in males is higher than in females with the ratio of males to females ranging from 1.4:1 to 2.4:1. Chordoma manifestations usually appear in the age range 50–60 years. These tumors can also appear at the age of 30–40 years, where 25% of these ages have chordomas on the spine and cranium [1-3].

Chordoma grows on the sacral foramina and blocks the nerve proximally. Posterior tumors tend to invade the muscles around the hip and the sacroiliac joints. Invasion to the anterior is rare due to the strong presacral fascia and the rectum is usually spared. Chordoma metastases occur in 30–40% of patients, where growth is slow and usually occurs after tumor recurrence even years after surgical resection. There have been secondary metastases in the lungs, bones, liver, lymph, soft tissues, and skin [2-4].

The therapeutic management of chordoma ranges from surgery, radiotherapy to chemotherapy. The main treatment options for chordoma management are surgery or resection [1,2,5]. The goal of surgery in the management of chordomas is to eradicate the tumor by maintaining as much function as possible. Chemotherapy is rarely chosen because it has poor sensitivity and is not suitable for sacral chordomas. However, chemotherapy is an option for metastatic chordomas due to its benefit to force the progression of the tumor.

We report the case of a 58-year-old male patient with sacral chordoma, where surgery as the mainstay therapy was not chosen and chemotherapy was given to the patient due to the limited facilities available in our hospital.

CASE REPORT

A 58-year-old male patient came with a chief complaint of a lump in the waist. The patient complained about the lump for 11 months before being admitted to the hospital. The lump was initially 3 cm in size, and then, it got bigger. Six months before, the patient went to a hospital and had a computed tomography (CT) scan examination. From here, the patient was referred to another hospital with complaints of numbness and tingling in both legs and the first lump sample was taken 5 months before. Then, the patient was taken a second lump sample 2 months before. After that, the patient complained of open surgery scars and could not feel the urge to urinate and defecate from the last month. The patient also complained that the legs felt increasingly weak and they could not walk. History of hypertension, diabetes mellitus, asthma, tumors, and malignancy was denied by the patient.

General examination showed that no abnormality and vitals were stable with a blood pressure of 135/84 mmHg, heart rate of 82/min, respiratory rate of 16/min, and temperature of 36.8°C. From the local examination of the sacral region, it was found that the mass was accompanied by ulcers and bleeding on inspection.

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On palpation, the mass is 17 cm × 15 cm × 4 cm in size, hard, and fixated (Fig. 1).

Sacral-vertebrae CT scan investigations concluded that the mass of the os sacral tumor with extensive bone destruction measuring approximately 13.17 cm × 13.70 cm × 14.9 cm in the form of macrolobulation invading the L5 intraspinal canal, pre-sacral space, and ischiorectal fossa pressing the sacroiliac joint bilaterally suspecting the diagnosis of chordoma clivus with differential diagnosis of osteosarcoma (Fig. 2). The additional scan was conducted to establish the diagnosis which concluded that a solid mass was destroying the bilateral os of the sacrum and infiltrates the bilateral gluteus maximus and cutis-subcutis supporting the diagnosis of sacral chordoma. Noticeably, bone metastases were seen in the corpus VL 3 and VL 5 (Fig. 3).

The results of histopathological sampling showed fragments of tumor tissue arranged in lobes with chondromyxoid areas consisting of pleomorphic and vacuole cytoplasm. The nucleus with fine chromatin showed physaliforus cells supporting the diagnosis of chordoma. Based on the history, radiological, and histopathological examinations, a final diagnosis of sacral chordoma with VL3 and VL5 bone metastasis was made.

The patient was managed with chemotherapy procedures with the epirubicin and paclitaxel cytotoxic drug regimens. The outcomes and side effects of the drug regimen were observed. Chemotherapy procedures were given 3 times, but did not show significant outcomes. Usually, radiotherapy is considered as the mainstay treatment for such tumors, but it has not been done in our case due to limited facilities. The patient is planned to be referred for radiotherapy to a center that has advanced radiotherapy facilities.

DISCUSSION

Chordomas arise from embryonic notochordal remnants along with the axial skeleton, can grow toward the sacral foramina, and obstruct the proximal nerve causing the complaints of numbness and tingling in both legs [6,7]. Due to this, our patient felt increasingly weak, was not able to walk, and felt the urge to urinate and defecate

Various examinations are carried out in an effort to establish a diagnosis of chordoma such as radiology and histology. In our case, the diagnosis was confirmed through a CT scan and histopathological investigation. CT scan imaging concluded sacral chordoma with VL3 and VL5 bone metastasis. CT scans are performed to assess the extent of the bone involvement or damage and to detect patterns of calcification within the lesion because CT has a better degree of accuracy than magnetic resonance imaging in delineating calcification and involvement of the bone osteolysis. CT is also useful in planning the reconstruction of the resistant osseous defect in tumors of the proximal sacrum [8,9]. Histopathological investigation conducted and showed fragments of tumor tissue arranged in lobes with chondromyxoid areas consisted of pleomorphic cells, and the nucleus showed physaliforus cells. These histopathological features are typical of dedifferentiated chordoma [1,3,4].

The management of the chordoma takes into account metastasis of the malignant tumor itself and the patient's health condition including the function of the kidneys, heart, and other organs. According to many chordoma guidelines, the mainstay therapy is surgery. The goal of surgery in the management of chordomas is to eradicate the tumor by maintaining as much function as possible. For patients where surgical intervention is inoperable or where the results of surgery have worsened the patient's condition, radiotherapy or chemotherapy were performed. A combination of surgery and radiotherapy is often done to maximize outcomes. Chemotherapy is rarely chosen because it has poor sensitivity and is not suitable for sacral chordomas. However, chemotherapy is an option for metastatic chordomas due to its benefit to force the progression of the tumor [5-7].

In this case, the chordoma had metastasized to VL3 and VL5, due to which the surgery was difficult, resulting in the use of a chemotherapy procedure to suppress the progression of the malignant tumor. This decision conforms to the National Comprehensive Cancer Network (NCCN) recommendation which says that for metastatic disease, chemotherapy, and/or surgical excision and/or radiation therapy and/or best supportive care can be an option in chordoma. The management of the chordoma takes into account metastasis of the malignant tumor itself and the patient's health condition. The regimen used was cytotoxic chemotherapy, such as epirubicin and paclitaxel [2,5-7].

Epirubicin as a derivative of doxorubicin has been shown to have anti-neoplastic effects when used alone or in combination. Besiroglu *et al*. have used epirubicin as a therapy for soft-tissue sarcoma and proved that those anticancer drugs have fewer cardiotoxicity effects than doxorubicin. Paclitaxel is a natural anticancer drug that has a unique mechanism of action used widely to treat cancer. Lee *et al*. reported a well-controlled case of metastatic chordoma using paclitaxel [10-13]. Chemotherapy procedures have been carried out 3 times, but there were no significant results in our study. During chemotherapy, the patient complains of urinary incontinence by means of inserting a urinary catheter and decreased inferior motor function. This may be due to the intrinsic chemo-resistance of the chordoma as stated by

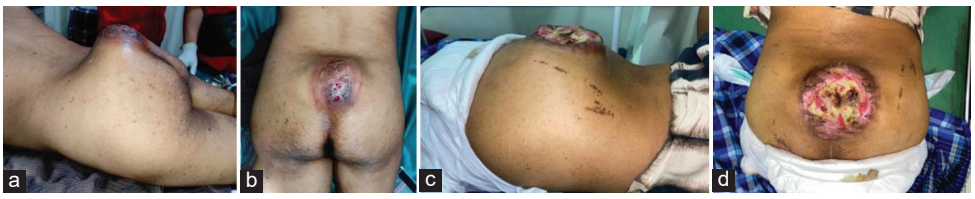

Figure 1: Lump in the waist (a) and (b) 6 months before; (c) and (d) At the time of admission to the hospital

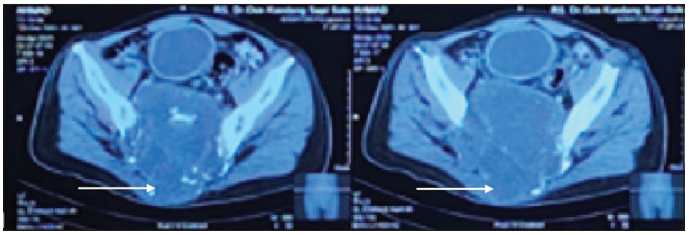

Figure 2: Computed tomography scan vertebrae sacral. White arrow depicting the sacral chordoma

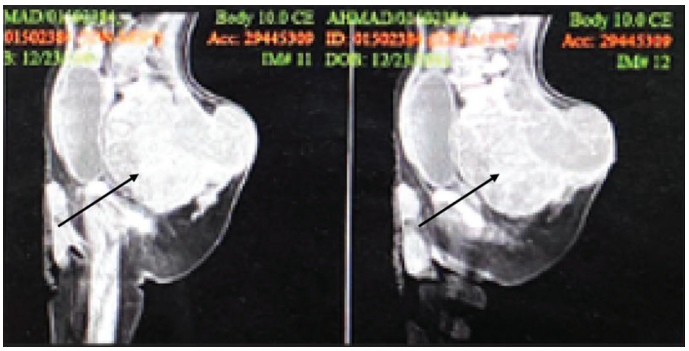

Figure 3: Additional computed tomography scan vertebrae sacral. Black arrow depicting the sacral chordoma

Verma *et al*. [14]. According to NCCN recommendation, radiation therapy can be the other option in chordoma management [5,7]. Radiotherapy has not been carried out in our patient due to limited facilities available and, hence, referred to another center for radiotherapy.

CONCLUSION

We presented the case of a 58-year-old male who was diagnosed with metastasis sacral chordoma. The diagnosis was confirmed with systemic anamnesis, physical examination, and supporting examination such as CT scan and histopathology. The management was chosen in accordance with the NCCN guidelines. Epirubicin and paclitaxel chemotherapy were carried out 3 times but did not show significant results due to the intrinsic chemoresistance nature of the chordoma. Radiotherapy is considered as the mainstay treatment but has not been done due to limited resources in our case and, hence, referred to a hospital with advanced radiotherapy facilities.

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