Challenges in Managing Locally Aggressive Pelvic Chondrosarcoma: A Case Report

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Abstract: Chondrosarcoma (CS) is a primary bone malignancy which differentiated from the cartilage tissue. It accounts for approximately 20% of malignant bone tumours. It ranks the third most common primary bone malignancy after osteosarcoma and myeloma. About 25% of the chondrosarcoma was reported in the pelvic region and the rest was reported in the periphery of the body. The management of pelvic chondrosarcoma is extremely challenging because it is radioresistant and chemoresistant. Therefore, surgical resection is the only curable treatment. However, completely surgical resection is impossible due to the close relationship between the tumour and the other pelvic structures. We report a case of locally aggressive pelvic chondrosarcoma in which sacrectomy was done. Bilateral iliac bone was fixed to the lumbar vertebrae by using screw fixation. However, the patient did not survive after he suffered from a number of post-operative complications.

Keywords: Pelvic chondrosarcoma, Primary bone malignancy, Osteosarcoma, Myeloma, Sacrectomy

INTRODUCTION

Chondrosarcoma (CS) is a malignant skeletal tumour with cartilaginous differentiation [1]. Chondrosarcoma accounts for approximately 20% of malignant bone tumours [2]. It is the third most common primary malignancy of bone after myeloma and osteosarcoma [2]. It is preferentially located in the pelvis in 22 to 39% of cases [1]. Compared to periphery chondrosarcoma, pelvic chondrosarcoma is more aggressive due to its location inside the pelvic cavity [3]. More than 70 per cent of the reported cases were found to have infiltrated to the adjacent structures such as bowel, bladder, nerve and vessels [3].

The principle treatment for chondrosarcoma is surgical resection because it is chemoresistant and radioresistant [1, 2]. However, pelvic chondrosarcoma is difficult to resect completely because of its close proximity to the surrounding vessels, nerves, soft tissues and organs in the pelvic cavity [1, 2]. Therefore, patient with pelvic chondrosarcoma has lower prognosis compared to patient with periphery chondrosarcoma [1, 3].

We report a case of locally aggressive pelvic chondrosarcoma in a young man who had no known medical illness and no family history of malignancy. Sacrectomy was done and bilateral iliac bone was fixed to the lumbar vertebrae by using screw fixation. However, the patient did not survive after he suffered from a number of post-operative complications.

Written informed consent was obtained from both the patient and his wife to discuss his case in the form of a published case report.

CASE REPORT

This is a case of a 34-year-old Malay male technician, an active smoker who started to smoke 20 years ago with a 30 pack-year history. He complained of progressive worsening of lower back and pelvic pain for 1 year and intolerable pain not relieved by oral analgesic for 1 week prior to admission. He had loss of appetite and loss of weight for 30kg within 1 year. The pain was associated with bilateral lower limb weakness and reduced sensation for 2 months prior to admission. Besides, he also complained of change in bowel habit such as constipation and urinary symptoms such as urgency, hesitancy and frequency.
On further questioning, he denied history of fever, night sweat and haemoptysis which could be symptoms of infection and tuberculosis. There were no previous history of trauma, family history of musculoskeletal, haematological, bowel and bladder malignancy and family history of congenital or degenerative bone diseases.

On physical examination, he appeared cachexic with body mass index (BMI) of 17kg/m² which was underweight. He was alert but in pain. His vital signs were stable with pulse rate of 80 beats per minute, respiratory rate of 16 breaths per minute, blood pressure of 110/70mmHg and he was afebrile. He was anemic and he had no jaundice. He was unable to stand and walk due to the pain. There was tenderness over lower lumbar and sacral region. Reduced motor power (4/5) and sensations (1/2) were noted over L2, L3, L4, L5 and S1 of the right side. There was also reduced sensation (1/2) over S2 and S3 of the right side. Anal tone (S4 and S5) was intact. Right peripheral nerves like sciatic, tibial and common peroneal nerves had reduced power (4/5) and sensations (1/2) as well. Per-abdominal examination revealed a tender, hard and smooth suprapubic mass extending to the right iliac fossa. The lower border of the mass was not felt and this suggested a pelvic mass. Examinations of other systems were normal.

Contrast-enhanced computed tomography (CECT) showed a heterogenous enhanced solid mass measuring around 10 x 8 cm in the pelvic cavity involving the right sacral S1, S2 and S3 right sacral ala. The mass extended into the central pelvic cavity anterior to the sacral and lower lumbar spines. This locally aggressive bony tumour mass displaced and compressed the adjacent small and large bowel and abutting the recto-sigmoid colon. There was no clear plane of demarcation between the mass and the pelvic soft tissues. No radiological signs suggestive of distal metastases such as lung and liver metastases.

Magnetic resonance imaging (MRI) of the abdomen and pelvic region showed a huge irregular mass in the pelvic cavity measuring about 10 x 8 x 12cm (AP x W x CC). It was isointense to the muscles on T1W1, slightly hyperintense on T2W1 and markedly hyperintense on T1RM, with central area of necrosis. The mass showed heterogenous enhancement following contrast injection except the central necrotic component which did not enhance. The craniocaudal extension of the mass was from the superior margin of L5 vertebral body to inferior margin of S5. Both CT and MRI suggested presence of locally aggressive pelvic tumour.

A heterogenous-enhanced solid mass (Arrow) was seen in the pelvic cavity involving right S1, S2 and S3 sacral ala. The mass extended into the central pelvic cavity anterior to the sacral and lower lumbar spine, L5. Right sacroiliac joint was invaded by the mass. Central hypodense area suggested central necrosis. The surrounding muscles and structures were infiltrated by the mass as well.

Full blood picture showed that he had normochromic normocytic anemia with Haemoglobin level of 8.1g/dL. Otherwise, the white cell count and platelets level were normal. Liver function test was normal except the alkaline phosphatase (ALP) level was elevated with value of 324U/L. Renal function and other blood tests were normal.

The patient was counselled and scheduled for operative management. The operation involved two
stages. The first stage involved the bilateral ilioinguinal incision to explore the pelvic cavity. This anterior approach aimed to visualise the mass and to know whether the mass has invaded structures like bladder, vessels and nerves. During the operation, the mass was found locally aggressive and has invaded the anterior abdominal wall and some neurovascular bundles. However, the bladder and bowel were spared. The mass was detached from the adjacent structures and became mobile. The ilioinguinal incisions were then sutured back.

The second stage of the surgery involved lower posterior vertical incision to approach the sacrum. The nerve roots and sacroiliac joint were carefully separated. The mass was then removed together with the sacrum. The mass was huge, measured around 13 x 10 cm with irregular margin and highly vascularised. Bilateral iliac bones were fixed to the lumbar bone by using screw fixation technique. The pelvic cavity was washed using normal saline and the wound was sutured. Post-operatively, he was given intravenous (IV) cefuroxime 750mg TDS and metronidazole 500mg TDS to prevent infection. He was sent to intensive care unit (ICU) and ventilated.

The challenges appeared 2 days after the operation. The patient was febrile and did not defecate for 2 days. He also had reduced urine output. Abdominal radiograph showed that the small bowel was dilated and suggested paralytic ileus. Blood culture and sensitivity isolated Pseudomonas aeroginosa. Arterial blood gas showed that he had severe metabolic acidosis (pH<7.1) and renal function test showed that he had disproportionate elevation of urea and creatinine which suggested acute kidney injury. The patient was hemodialysed and given broad spectrum antibiotics such as IV meropenem 1g TDS and IV penicillin 2 million units QID to treat the sepsis. He was monitored tightly in the ICU. However, he passed away 1 week later after he suffered from multi-organ failure, sepsis, severe metabolic acidosis, acute kidney injury and acute respiratory distress syndrome.

The biopsy of the pelvic tumour showed grade I chondrosarcoma with mild hypercellularity. The chondrocytes had plump vesicular nuclei with small nucleoli. Binucleate cells were sparse and mitotic figures were almost absent.

**DISCUSSION**

Pelvic chondrosarcoma is a very challenging orthopaedic oncology disease. The challenges can be divided into intra-operative and post-operative challenges [1, 4]. These challenges will directly and indirectly influence the prognosis of the patient.

Intra-operative challenge mainly refers to difficulty in getting a clear surgical resection margin [1, 2, 4]. Intra-operative margins had been classified as ‘wide’, ‘wide contaminated’, ‘marginal’ and ‘intralesion’. Only wide margins had been considered to be adequate and contaminated margins had been regarded as positive. The criteria for contaminated margins had been related to the operative description of the surgeon. If the tumour had been violated during resection, even if additional tissue had been removed for achieving wider margins, then the margins had been considered to be wide contaminated. The final decision about margins had been only achieved after obtaining a consensus among surgeons, and reviewing of the pathology [4]. Wide resection is impossible in some cases due to the aggressiveness of the tumour which has invaded the adjacent important structures [1, 3, 4]. This creates a challenge to the surgeon in this case because the tumour has infiltrated the neurovascular bundle and wide resection will compromise blood supply to the lower gastrointestinal and genitourinary tract, as well as lower limb and sexual organ [3, 4].

The prognosis of the chondrosarcoma depends mainly on its staging, grading and location. A study has shown that histological grade is the single most important univariate and multivariate predictor of the overall survival of the patients; the higher the histological grade, the lower the survival to death, higher local recurrence and metastasis [2]. Therefore, if wide local resection is impossible and the tumour is a high grade tumour, the local recurrence rate will be very high and long term prognosis will be very poor.

The common post-operative complications of the pelvic surgery include infection, bleeding, paralytic
ileus, neurovascular injury, incontinence and sexual dysfunction [5, 6]. Among these complications, infection is the most common complication [1, 5]. Infection is disastrous in the case of post pelvic operation because the area is highly vascularised and any foci of infection will promote systemic sepsis and multi-organ failure [5]. This complication is the one being encountered in this case. Paralytic ileus and intestinal obstruction worsen the sepsis by promoting bacterial translocation from the gastrointestinal tract [7]. Cancer patient has generally lower immunity [8] and take all these points into consideration, post-operative infection is difficult to control despite optimum antibiotics treatment.

CONCLUSION

Histological grade was the single most important univariate and multivariate predictor of the overall survival of the patients; the higher the histological grade, the lower the survival to death, local recurrence and metastasis.

Pelvic chondrosarcoma is difficult to treat and many challenges will be faced throughout the management. Wide local resection is the main challenge being faced intra-operatively and infection is the most common post-operative challenge. Since chondrosarcoma is radioresistant and chemoresistant, new method such as immunotherapy, biologic therapy or other method should be studied to find out whether there is any other method which can treat chondrosarcoma apart from surgery as wide resection is not easy in the case of pelvic chondrosarcoma. As for the post-operative infection, the only way to reduce the incidence is to provide sufficient prophylactic and post-operative antibiotic dose and maintain highly-sterile operating environment.

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