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GIANT CELL TUMOR OF SACRUM A REPORT OF TWO CASES

SAKRUMUN DEV HÜCRELI TÜMÖRÜ İKİ OLGU SUNUMU

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SUMMARY:

Giant cell tumor (GCT) of bone is a rare neoplasm that accounts for approximately 5% of all primary bone tumors in adults. Sacrum is an unusual site for giant cell tumor and it is an intriguing and unpredictable entity with a high predilection for recurrences. We hereby report 2 cases of Giant cell tumor of sacrum occurring in a 42- year- old male and a 17- year- old female. The tumor recurred twice in the 42- year- old male and was treated with surgery radiotherapy and embolization, whereas the tumor did not recur in the 17- year- old female who was operated for the same.

Key words: Giant cell tumor, sacrum, surgical treatment

Level of evidence: Case report, Level IV

ÖZET

Kemiğin dev hücreli bir tümör olup primer kemik tümörlerinin % 5'ini oluşturur. Sakrum sık tutulan bir bölge değildir buna karşın tanısı zor ve nüks riski daha yüksektir. Bu çalışmada sakral dev hücreli tümörü olan biri 17 yaşında kadın, diğeri 42 yaşında erkek hasta sunulmuştur. 42 yaşındaki erkek hasta tümör eksizyonunu takiben iki kez nüks etmiş ve radyoterapi ve embolizasyon uygulanmıştır. Diğer taraftan 17 yaşındaki kadın hasta aynı zamanda opere edilmesine rağmen nüks görülmemiştir.

Anahtar Kelimeler: Dev hücreli tümör, sakrum, cerrahi tedavi

Kanıt Düzeyi: Olgu sunumu, Düzey IV

INTRODUCTION:

Giant cell tumor (GCT) of bone is a rare neoplasm that accounts for approximately 5 % of all primary bone tumors in adults[9]. GCT most frequently occurs at the ends of long bones, sacrum is an uncommon site[11]. In all locations the neoplasm occurs most commonly between the ages of 25-40 years and it affects males and females with equal frequency[1]. Various treatment methods have been advocated including arterial embolization, curettage, surgical excision, radiation and cryotherapy [8]. Treatment is very successful in long bone lesions, but the optimal treatment and medical management of GCT in spine and sacrum has not been well established[8].

We hereby describe two cases of Giant cell tumor of sacrum, one occurring in a 17 year old female and another in a 42 year old male.

CASE REPORTS:

Case No.-1:

A 42 years-old-male reported to our Neurosurgical OPD in 2009 with complaints of pain and weakness of both lower limbs. On examination power in the lower limbs was 4/5 proximally and 3/5 distally. There was sensory deficit along L5 - S1 dermatomes. MRI spine showed a mass in sacral area with bilateral paravertebral extension, involvement of bilateral sacroiliac joints with extension to spinal canal. Surgery was done and intraoperatively a soft friable highly vascular tumor involving L5-S3 extending laterally to paraspinal tissue (more on left side) compressing S1 nerve roots was seen. Bilateral dural sac was displaced posteriorly. His post-operative period was uneventful. He was subjected to radiotherapy (25 cycles). He was doing well till 2011 when he again started with weakness of both limbs. MRI has done showed recurrence of GCT. Patient was treated with embolization (bilateral ileolumbar branches) followed by re-exploration and excision of the lesion. Patient on a follow-up period of 3-yrs is doing well and is recurrence free (Figure-1).

Case No.-2:

A 17-years- old female came to the neurosurgical OPD with chief complaints of low backache and pain in both lower limbs for the past 1 year. There was no history suggestive of motor or sensory deficits. MRI showed an expansible destructive lesion in the sacrococcygeal area which was thought as chordoma. Surgery was performed, presacral, grayish, non suckable highly vascular tumor was removed. The sacrum was eroded and tumor was engulfing the nerve roots. Rectum was free. Her post-operative period was uneventful. On a 2-year follow-up period she is doing well.

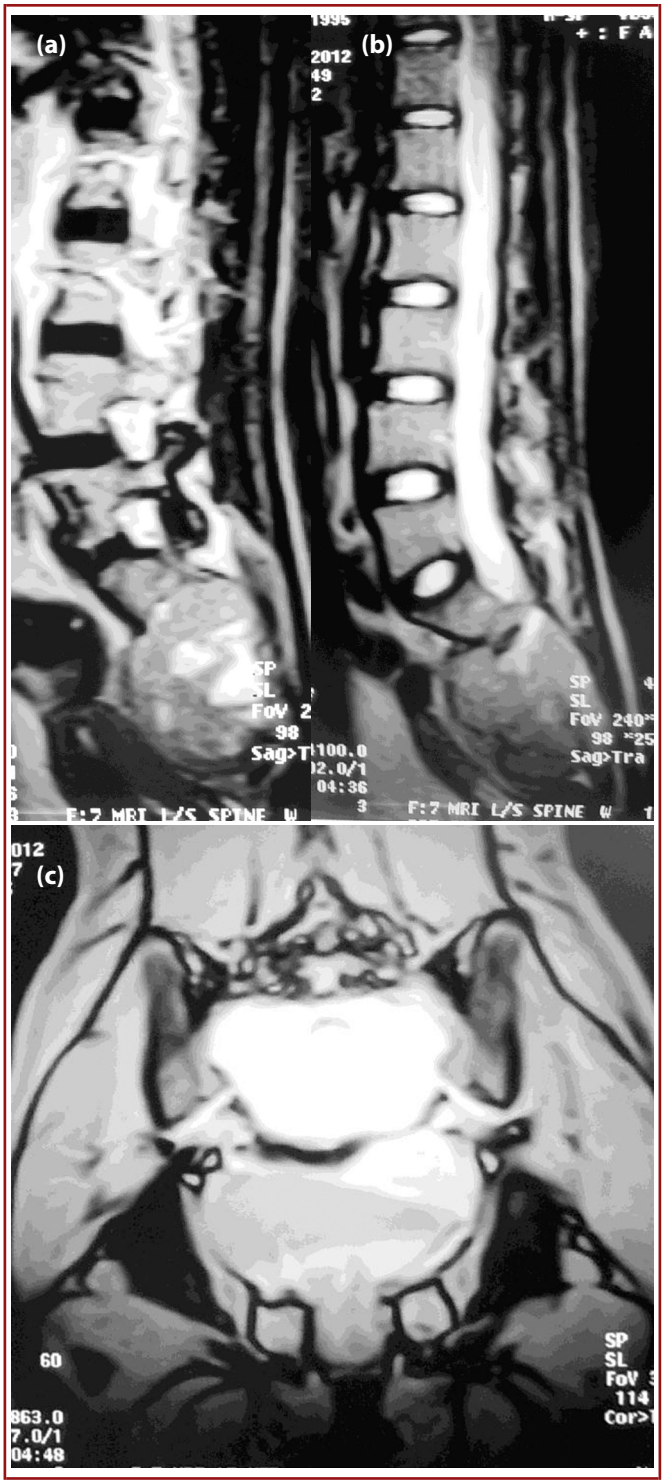

Figure-1. MRI lumbo-sacral spine sagittal sections T1 **(a)** and T2 **(b)** weighted images and **(c)** frontal MR view show a mixed intensity lesion involving the sacrum.

DISCUSSION:

Giant cell tumor of bone is a distinct neoplasm with some unusual behavioral characteristics. In an analysis of 10 cases of GCT sacrum by Martin et al[8], the mean age was 31 years with

an age range of 13-49 years. Our patients fell well within the age range, one being 17-years of age and the other 42-years. All his patients presented with pain for an average duration of 30-months and the pain most frequently radiated to the back and into the thighs. Both of our patients also presented with pain radiating to both lower limbs. Bladder and bowel disturbance however seen in 70% of his patients was not seen in our patients.

Kanamori reported a patient of giant cell tumor of sacrum who presented with low back pain and numbness on the posterior aspect of left thigh[5]. Liang also presented three case reports of giant cell tumor of sacrum in a 31-year-old male and two females' aged 1- yr and 31-yrs[7]. In the cases analyzed by Martinet al[8], the sacral lesions were large, poorly defined lytic masses arising in the central part of the sacrum and spreading to involve both wings of the sacrum. Three patients in his series had an associated soft tissue mass. The most frequent location was the upper two segments of the sacrum, although several lesions were so large that they involved the entirety of the sacrum. We also experienced predominantly lytic lesions involving the entire sacrum in both the cases.

Kanamori in a study on a recurrent sacral GCT noted a large lytic and destructive bone lesion below the level of the first sacral segment[5]. No calcifications were noted within the mass. MRI in his case demonstrated a sacral lesion which was of mixed intensity on T1 and T2 weighted sequences. Einestein also reported a large destructive lesion involving the distal half of the sacrum with extension to the sacrococcygeal joint. The superior margin of the lesion was poorly defined and there was no sclerosis at the edge of the lesion[3].

The gross appearance of a giant cell tumor is usually quite characteristic. The lesion is soft and dark brown. Microscopically, giant cell tumors consist of multinucleated giant cells and mononuclear cells. The giant cells are usually distributed uniformly throughout the lesion.

The giant cells may contain as few as 10 or as many as 50 nuclei. The mononuclear cells are round to oval, and the nuclei resemble those in the giant cells. Although mitotic activity may be brisk in the mononuclear cells, the nuclei lack atypia. Atypical mitotic figures should not be seen in a classic giant cell tumor.

Although most giant cell tumors have the classic appearance previously described, variations occur. It is not uncommon to see areas of secondary aneurysmal bone cyst in giant cell tumors. They may be focal or dominant. Collections of foam cells are quite common in giant cell tumors. The mononuclear cells tend to spindle out in these foam cell areas and may even have a storiform pattern. Occasionally, the spindling cells with the storiform pattern dominate the appearance of the giant cell tumor itself (Figure-2)[10].

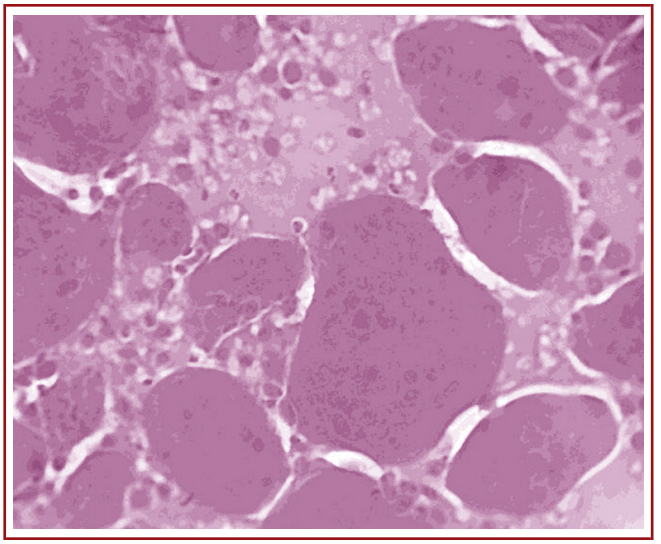

Figure-2. Photomicrograph showing multinucleate giant cells in Giant cell tumor (Squash smears Haematoxylin and eosin x 40)

The differential diagnosis of giant cell tumors includes a variety of conditions associated with giant cells, including aneurysmal bone cysts, osteosarcomas, hyperparathyroidism, and chondroblastomas. Calcification or chondroid differentiation should separate chondroblastoma from a giant cell tumor. Most other giant cell-containing lesions are metaphyseal or diaphyseal, whereas giant cell tumors tend to be epiphyseal. Giant cell tumors have been shown to express p63, suggesting that this may be a useful biomarker to differentiate giant cell tumor from other giant cell-rich tumors[2].

Histological features in our case were typical of giant cell tumor of the bone. The bone showed haemorrhage, reactive bone formation, admixed with many giant cells. There was no significant pleomorphism to lead to the consideration of malignancy. Our first patient was treated with resection and he had no residual neurological deficits at follow up.

Intralesional resection of sacral GCT is associated with lower post- operative morbidity than sacrectomy[4,6]. Giant cell tumor of bone remains a difficult and challenging management problem because there are no absolute clinical, radiographic or histological parameters that accurately predict the tendency of any single lesion to recur[7].

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