« Case Report»

Primary Ewing sarcoma of the sacrum

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Abstract

Primary malignant sarcomas of the spine are extremely rare. Because of biological heterogeneity, these tumors have variable sensitivity to radiation and chemotherapy. Adequate local control through complete tumor removal is an important therapeutic goal. However, aggressive resection of tumors in the spinal column must be coupled with restoration of spinal column stability and minimization of neural deficits. The balance of these factors makes treatment of primary sarcomas of the spine challenging, and dictates an individual approach to treatment. We report a 17 years old lady with chief compliant of low back pain and saddle anesthesia the clinical picture and imaging characteristics were analyzed as well as the management modalities and outcome.

keywords: Ewing´s sarcoma, primary spine tumor

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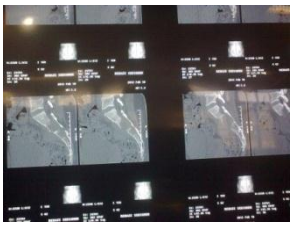
Introduction

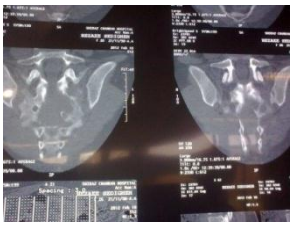
Primary malignant sarcomas of the spine are extremely rare. It has been estimated that they account for only 3.5% to 14.9% of all primary bone sarcomas 1-5. In the study of primary vertebral Ewing's sarcoma ES), the division of the spine into nonsacral and (sacral is important and is dictated by the diff e re n t behavior of ES in these two regions in terms of response therapy and survival rates (2, 3, 4, 15, 8, 7, 12, 14). Most studies on ES involving the mobile spine are limited to case reports. A few larger series evaluated patients with primary ES affecting the mobile and nonmobile spine (1, 2, 3, 4, 6). Microscopic examination demonstrates small, round blue cells with a uniform appearance. Electron microscopy and cytogenetic studies help differentiate ES from other small, round blue cell tumors. The translocation t (11; 22) (q24; q12) is identified in most cases of ES, 7. Adequate treatment of primary malignant spine tumors is aimed at cure rather than palliation whenever possible. The definitive management of Ewing's sarcoma of the spine, as in other locations, could include three main modalities: surgery, radiotherapy, and combination chemotherapy, 1 the purpose of the current study is to report the diagnosis and the

Case

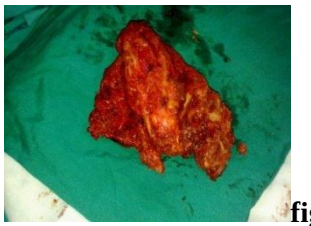
A 17 years old lady with chief compliant of low back pain and saddle anesthesia.in neurological examination and no motor deficit was detected and patient ambulated. The pain was intractable and not response to analgesics for 3 months, Sensibility was decreased, the results of laboratory studies, including erythrocyte sedimentation rate were within normal limits. Computed tomographic (CT) (fig1, 2) and magnetic resonance imaging (MRI) evaluation of the patient showed a presentation as determined soft tissue mass invading the epidural space . The patient was without evidence of metastatic disease by staging chest radiographs, chest computed tomography scans and bone scans. The first line of management by surgical planning was performed for patient and she underwent enblock resection of the sacrum (fig3, 4).the specimen was sent for pathologic examination and was reported Ewing sarcoma of the sacrum. After surgery she could ambulated but was incontinent.



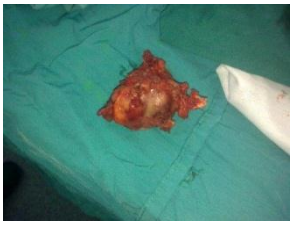
fig,1



fig,2



fig,3



fig,4

Discussion

Ewing sarcoma is a primitive neuroectodermal tumor originating from the medullary cavity of the long bones often arising in the diaphysis or medullary-diaphyseal portion and occurring in the group of 10-30-year-old males (1, 8). ES is a rare entity and only a few cases have been reported in the literature5. In 1991, Kaspers et al. (5) firstly reviewed 15 cases of ES and presented the clinical features. However, there were still no reports about the treatment and prognosis of the epidural Ewing sarcoma. We reviewed 9 reported cases with our one additional patient2, (4), (9), (14), (15). Since combined radiotherapy and chemotherapy are effective for Ewing's sarcoma, extensive laminectomy could cause scoliosis or spinal deformities in children. Needle biopsy for the diagnosis is recommended. In our case, marked spinal cord compression with symptoms of progressive neurological deficits was noted, so we performed complete resection of the tumor for immediate decompression. Postoperatively, the patient demonstrated rapid recovery of her neurological deficits. The mean age was 20.4 years with male predominance. The gender ratio of female: male was 1: 2.3. Lumbar spine (50%) followed by thoracic spine (30%) and cervical spine (10%) were the most common sites in patients with ES. Symptoms and signs depended on the tumor location and the severity of spinal cord compression. Painful sensation (100%) followed by weakness of limbs (70%) and numbness of extremities (50%) were the most common clinical presentations. Regarding location of pain, 11 of20 patients (55%) complained of backache (3, 7). The time interval between initial clinical presentations to surgical intervention varied from one to 18 months, with a mean of five months. According to the surgical intervention, seven patients (35%) underwent total removal of tumor, eight patients (40%) underwent partial tumor resection, and the nearly 25% others had undetermined surgery. With respect to outcome, 10 patients died during a mean follow-up time of 16 months (1-48 months) and most patients (80%) had lumbosacral ES. Comparing survival, 71% of patients who underwent complete resection of the tumor with adjuvant therapy were free of disease. In contrast, only 25% of cases who had partial tumor resection were disease-free. Complete tumor resection with adjuvant radiotherapy and chemotherapy seemed to be more beneficial for improving prognosis (5, 8, 9). Tumor location may be one of the prognostic factors of the Ewing sarcoma, but the issue is still controversial1. In our review, cases with ES occurring in the lumbosacral spine had a lower survival rate and highly metastatic tendency compared to cases with cervicothoracic localization. It seemed that the prognosis of ES is much poorer over the lower spinal region, but it was still hard to draw any conclusion because of the limited number of cases. A possible explanation for the difference in prognosis may be that the larger epidural space in the lower-third of the spine delays the presentation of symptoms and the diagnosis of the cancer. In the evaluation of an epidural spinal tumor, it is important to distinguish ES from meningioma, neurofibroma, dermoid cyst, abscess, embryonal rhabdomyosarcoma, chondrosarcoma schwannoma, synovial sarcoma, osteogenic sarcoma, hemangioma, nerve sheath tumor, lymphoma, and leukemia (2, 5, 8, 11, 15, 9). MR imaging, superior to myelography and computed tomography, is considered to be the best diagnostic method to delineate the spinal disorders. On MR images, ES is generally of low-to-isointense signal compared with muscle on T1-weighted images, of high signal intensity on T2-weighted images, and

exhibits heterogeneous enhancement2. However, it is difficult to differentiate between ES and Immunohistochemical neoplasms based on clinical, radiological and laboratory examination. Staining such as immunoreactivity to glycoprotein p30/32 (CD99) and ultrastructural and molecular biological technique could be helpful in establishing the diagnosis1. In conclusion, ES is a rare and aggressive neoplasm. Early decompressive surgery is strictly indicated to secure the vital neurological functions. Complete tumor resection with adequate radiotherapy and chemotherapy is considered as the optimal therapeutic policy. However, due to the limited number of reported cases, an accumulation of such cases is needed for further evaluation and research to prove its histogenesis.

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References

1-Sharafuddin MJ, Haddad FS, Hitchon PW, et al. Treatment options in primary Ewing's sarcoma of the spine: report of seven cases and re view of the literature. Neurosurgery 1992;30:610-618.
2-Bacci G, Picci P, Gherlinzoni F, et al. Localized Ewing's sarcoma of bone: ten years' experience at the Instituto Ortopedico Rizzoli in 124 cases treated with multimodal therapy. Eur J Cancer Clin Oncol 1985;21:
3-Klaassen MA, Hoffman G. Ewing's sarcoma presenting as spondylolisthesis. Report of a case. J Bone Joint Surg 1987;69:1089-1092.
4-Pilepich MV, Vietti TJ, Nesbit ME, et al. Ewing's sarcoma of the vertebral column. Int J Radiat Oncol Biol Phys 1981;7:27-31.
5-Wilkins RM, Pritchard DJ, Burgert EO, Unni KK. Ewing's sarcoma of bone: experience with 140 patients. Cancer 1986; 58:2551-2555.
6-Barbieri E, Frezza G, Martelli O, et al. Non conventional fractionation in radiotherapy of the musculo-skeletal sarcomas. Tumori 1998;84: 167-170.
7-G rubb MR, Currier BL, Pritchard DJ, et al. Primary Ewing's sarc o m a of the spine. Spine 1994;19:309-313.63-173.
8-Harimaya K, Oda Y, Matsuda S, Tanaka K, Chuman H, Iwamoto Y. Primitive neuroectodermal tumor and extraskeletal Ewing sarcoma arising primarily around the spinal column: report of four cases and a review of the literature. Spine 2003; 28: E408-E412.
9-Shin JH, Lee HK, Rhim SC, Cho KJ, Choi CG, Suh DC. Spinal epidural extraskeletal Ewing sarcoma: MR findings in two cases. AJNR Am J Neuroradiol 2001; 22: 795-798.
10-Angervall L, Enzinger FM. Extraskeletal neoplasm resembling Ewing's sarcoma. Cancer 1975; 36: 240-251.
11-Gandhi D, Goyal M, Belanger E, Modha A, Wolffe J, Miller W. Primary epidural Ewing's sarcoma: case report and review of literature. Can Assoc Radiol J 2003; 54: 109-113.
12-Kaspers GJ, Kamphorst W, van de Graaff M, van Alphen HA, Veerman AJ. Primary spinal epidural extraosseous Ewing's sarcoma. Cancer 1991; 68: 648-654.
13-Venkateswaran L, Rodriguez-Galindo C, Merchant TE, Poquette CA, Rao BN, Pappo AS. Primary Ewing tumor of the vertebrae: clinical characteristics, prognostic factors, and outcome. Med Pediatr Oncol 2001; 37: 30-35.
14-Tefft M, Vawter GF, Mitus A. Paravertebral "round cell" tumors in children. Radiology 1969; 92: 1501-1509.
15-Angervall L, Enzinger FM. Extraskeletal neoplasm resembling Ewing's sarcoma. Cancer 1975; 36: 240-251.
16-Mahoney JP, Ballinger WE Jr, Alexander RW. Socalled extraskeletal Ewing's sarcoma. Report of a case with ultrastructural analysis. Am J Clin Pathol 1978; 70: 926-931.