PRIMARY EWING SARCOMA OF THE SPINE: REPORT OF TWO CASES

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Ewing sarcoma is a primary bone tumor which was originally described by Lucke in 1866 but was defined in such detail by Sir James Ewing in 1921 that it is now associated with his name [1]. This tumor involves the spine primarily in 3.5% to 14.9% of cases [2,3]. The review of 22 cases of Ewing sarcoma of the spine by Pilepich et al in 1981 [4] remains the largest study of this pathology in such location in the literature and apart from a few smaller series [3,5], most of the documentation of Ewing sarcoma of the spine has been by isolated case reports [6-8].

Considering the worldwide rarity of primary spinal Ewing sarcoma, we were surprised to encounter two cases among the 30 cases of spinal tumors treated in our unit over the last seven years [9]. There are a number of articles reflecting on the experience with Ewing sarcoma in Saudi Arabia [10-12]. However, to our knowledge, in none of these reports was the spine the primary site of the tumor. We feel justified, therefore, in presenting our two cases to highlight awareness of clinicians to the occasional involvement of the spine as a primary site of Ewing sarcoma, which should be considered in the differential diagnosis of spinal tumors causing cord compression.

Case Histories

The clinical characteristics of the two cases appear in the Table (Figures 1-3). The chest x-ray, abdominal ultrasound, skeletal survey and isotope bone scans for both cases were normal. The histology in both cases (Figures 4 and 5) was distinct nuclear membrane and a finely divided chromatin with a single minute nucleolus. The cytoplasm was ill defined, pale staining, with large vacuoles full of glycogen. Electron microscopy in Case 1 showed the cells possessing rounded nuclei with some cleaving, multiple minute nucleoli and small amounts of chromatin. The cytoplasm contained abundant glycogen as well as occasional

FIGURE 1. CT scan with intrathecal contrast (Case 1) shows right-sided erosion of lamina of S1 and extradural mass (arrow heads) displacing theca to left.

FIGURE 2. CT scan (Case 1) shows the eroded sacrum and the pelvic tumor mass (arrow heads).
Discussion

Ewing sarcoma frequently affects patients in the second decade with an increased incidence in males above the age of 13. It is considered to be very rare below the age of five years and above the age of 30 years [1]. Both of our cases were males; Case 2 was 11 months of age at the time of diagnosis, which is very unusual [1].

Our radiological investigations were limited to plain x-rays, myelography, CT scans and isotope bone scans. The latter were performed to exclude other tumor foci since Ewing sarcoma is frequently multifocal and up to 25% to 50% of cases may have disseminated disease at presentation [1]. Neither of our cases had any evidence of metastasis and their tumors were primarily spinal. The plain spinal x-rays were sensitive in demonstrating the bony erosion in Case 1 but not in Case 2, even though at operation there was some evidence of infiltration of the lamina by tumor, which had also involved the paraspinal muscles. It is recognized that the changes in the plain x-rays may appear late [3,13]. For both cases, the isotope bone scans were performed after surgery and were found to be normal. It is reported, however, that in Ewing sarcoma, isotope bone scans are useful in identifying the primary lesion and metastasis [3,14]. Nowadays, MRI is the most sensitive imaging modality in the detection of Ewing sarcoma of the spine [15]. It is valuable in defining the surgical margin of the tumor and for monitoring the response to chemotherapy. Both of our cases were diagnosed before the availability of immunocytochemical techniques in our hospital. The light and electron microscopic findings in our cases, the location of their tumors and their clinical features were supportive of the diagnosis of Ewing sarcoma. It has been reported recently that cytogenetic analysis can be a valuable tool in the diagnosis of Ewing sarcoma and that the finding of a specific chromosomal abnormality may be the only clue in establishing the diagnosis when other criteria are not conclusive [16].

Since most of spinal Ewing sarcomas invade the canal from behind, a laminectomy is commonly the approach of choice [1,3]. Both of our cases had a decompressive laminectomy because of deteriorating neurological condition. In addition, both patients were treated with dexamethasone (dose 8-12 mg/day) followed by a tapering dose for seven days, which is considered beneficial in the early management of all patients with spinal tumors [17].

*VAC=vincristine, adriamycin and cyclophosphamide.
It is recognized that for Ewing sarcoma of the spine, surgery is indicated in the presence of extradural compression with neurological deficits, for stabilization of the spine, following a poor response to an initial primary chemotherapy or radiotherapy treatment, to treat residual disease and for sacral tumors which are recognized to have a more aggressive biological behavior and a poorer prognosis [2-4]. Recently, trials with initial chemotherapy produced a rapid extradural decompression in a large number of cases [17,18]. It is therefore recommended that in the neurologically stable patient, initial chemotherapy after biopsy should be administered to the primary tumor before the definitive local treatment by surgery or radiotherapy is instituted [3,17,18]. Both of our cases were treated with the VAC regime. Various regimes are being tried now and it appears that the high-dose intermittent regime of vincristine and doxorubicin alternating every three weeks with vincristine and cyclophosphamide is likely to provide the best results [19].

The prognosis of Ewing sarcoma has improved tremendously in the last 20 years following the use of chemotherapy. It is recognized that sacral tumors are associated with a worse outcome because of their location and they are more likely to be advanced at the time of diagnosis in addition to the technical difficulty in supplying adequate radiation or achieving total surgical excision. In the intergroup Ewing sarcoma study (IESS) experience, nonsacral spine lesions had 100% local control rate and 86% long-term survival rate while for sacral spine tumors, the local control rate was 62.5% and the long-term survival rate was 25% [4]. Other less favorable reports indicate that the five to 10 year survival rate for nonsacral spinal Ewing sarcoma is 25% to 29% [2,3]. One of our cases, whose tumor had involved the sacrum, had survived free of symptoms for seven years while the other case was followed up for only two years.

References


