Ewing's sarcoma

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Abstract: Ewing's sarcoma is a rare malignant neoplasm of bone marrow and its exact nature is unknown. It arises in the middle of a long bone and may produce metastases in other bones and lymph glands [1]. A 35 year old male presented with Ewing's sarcoma of the left clavicle that had metastasized to his cervical and thoracic spine. He had spinal cord compression for 5 years and had been treated for this condition. This case report covers his clinical history and management. The aetiology, epidemiology, radiographic appearances and clinical presentation of this disease are discussed [1-9].
Key words: Pancoast tumour, Horner's syndrome, skeletal metastases, bone tumour.

Case report

Following a motor vehicle accident five years ago a 35 year old male was incidentally diagnosed with Horner's syndrome and a pancoast tumour. At that time he underwent a computed tomography scan to exclude traumatic causes; the findings revealed a large left apical mass lesion. His previous surgical history included surgery of the ear, laminectomy and decompression surgery to the cervical and thoracic spine.

He recently presented with weakness in his left arm, pain, swelling, loss of weight and paraplegia for six months. A whole body bone scan was performed which revealed an irregular abnormal uptake of activity in the lower cervical and upper thoracic spine. An increased activity in the second, anterior left rib was noted but was considered to be non-specific. There was no evidence indicating skeletal metastases. Two days after the whole body bone scan was performed the patient underwent a computed tomography scan of the chest, abdomen and pelvis. The mass of the left apex had increased significantly in size since the previous scan done two years ago. Bony destruction of the adjacent rib, the left lateral aspect of adjacent vertebral bodies and transverse processes of the left posterior vertebral elements were visualized. There was definite involvement of bone marrow. Histology findings indicated the presence of an Ewing's sarcoma in the existing lesion rather than a small cell carcinoma. He was referred to the radiology department by the oncologists for further investigation.

A magnetic resonance imaging scan of his chest and vertebral column was performed and a large, in-homogenous, destructive mass in the region of the thoracic inlet/ left lung apex, lower cervical spine and upper thoracic spine region were visualised (see Figure 1). The origin of this large soft tissue mass, which measured 109 x 155 x 92 mm, was not clear due to the extensive and destructive soft tissue component. There was evidence of vertebral body collapse and destruction from the fifth cervical vertebra to the third thoracic vertebra (C5- T3). An extensive abnormal signal in the marrow of these vertebral bodies was evident. Abnormal soft tissue, related

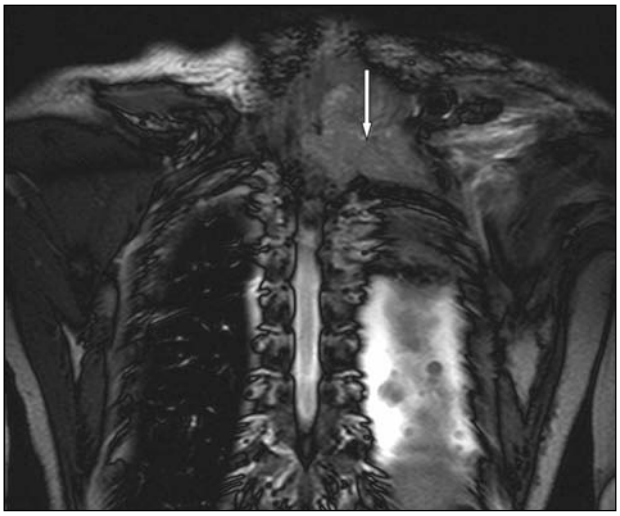

Figure 1: This T2 Coronal MR image visualizes a large, inhomogeneous, destructive mass in the region of the left lung apex (see arrow).

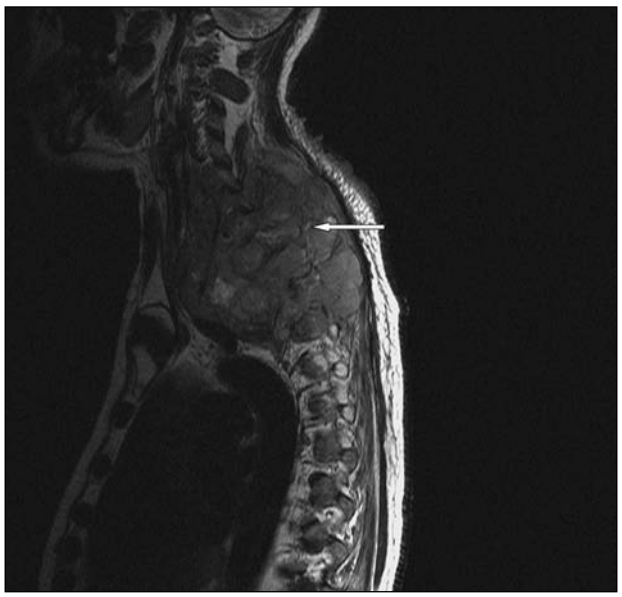

Figure 2: T2 sagittal MR image reveals the large tumour with accompanying multiple vertebral body collapse as well as destruction involving C5- T3 (see arrow).

to the posterior elements of these vertebral bodies, was noted. Prevertebral abnormal soft tissue was evident from C4-T2. There was spinal cord compression at the level of C6-T4 with extensive abnormal signal in the spinal cord from C3-T3. No abnormalities were found in the vertebral bodies of the lumbosacral spine (see Figure 2).

Discussion

Ewing's sarcoma represents approximately 7% of all primary malignant bone tumours and is the fourth most common primary malignant bone tumour [2]. The aetiology of Ewing's sarcoma is unknown; it arises from the medullary canal and involves the bone more diffusely, giving rise to uniform thickening of the bone [3]. There is no recognised agent but a specific chromosomal translocation in Ewing's sarcoma has been demonstrated between chromosomes 11 and 22; other similar translocations between chromosome 22, 21 and 17 have also been reported [4].

This neoplasm occurs in younger age groups compared to any other primary malignant bone neoplasm, usually between the ages of 5 to 15 years and rarely after the age of 30 [5]. This patient is older than 30 years which is considered to be out of the norm. Males are affected more frequently than females, with a male to female ratio of approximately 1.5:1 [5]. Patients in the younger age range usually have lesions in the peripheral skeleton, while older patients present with axial lesions [2]. Patients usually present with localized pain, tenderness and swelling in the region of the involved bone. There is frequently associated pyrexia [2]. The swelling is often red and hot and there may be an audible bruit. Thinning of the periosteum may result in a characteristic crackling on palpitation and there may be crepitus if a fracture has occurred. There is sometimes a history of preceding trauma, although no casual relationship exists [4].

Limb sparing operations could reduce the need for many patients to undergo limb amputation. Chemotherapy is given prior to surgery to induce tumour sterilisation and reduce tumour size. Limb salvage surgery is possible if an adequate surgical margin can be obtained while preserving enough tissue to maintain a functional limb [7]. Surgery is recommended as treatment of choice for lower extremity lesions in children with un-fused epiphyses [6]. Improvements of treatment results have occurred with chemotherapy given in combination with surgery and/or radiotherapy to the primary [7]. Chemotherapy may be as short as 6 treatments at 3 week cycles; however most patients will undergo chemotherapy for 6-12 months. Ewing's sarcoma is a radiosensitive tumour and radiation therapy has been the standard treatment of the primary site for many years. The entire affected bone must be irradiated because of the tendency of the tumour to permeate the marrow cavity [7-9]. Radiation therapy alone is reported to result in a local control rate of between 65% and 75%. After chemotherapy is added to adequate irradiation, this figure may rise to between 90% and 95%.[6].

Cytotoxic drugs are now also improving the outlook. The optimum regime is intensive radiation to heal the local lesion with concurrent chemotherapy to deal with microscopic secondary deposits.

Radiographically, the lesion is usually seen as a diffuse, destructive 'moth eaten' diaphyseal tumour. Extension to soft tissue is common and there is often a periosteal reaction which produces an 'onion skin' appearance [7].

Conclusion

The patient in this case report had decompression surgery to his cervical and thoracic spine. He has received his 5th cycle of chemotherapy in conjunction with pain alleviating drugs. In addition to this he has also been receiving physiotherapy of his upper and lower limbs.

Being diagnosed with Ewing's sarcoma should not let a patient lose hope of recovery. Increasingly effective chemotherapy and surgery have improved the prognosis to an 80% five year survival rate. Magnetic resonance imaging is the best imaging modality to demonstrate the exact extent of a mass. The high quality of soft tissue detail obtained with this imaging modality enhances tumour detection and subsequent treatment thereof.

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