Ewing’s sarcoma

Monique C Lewis 3rd year student radiographer (Diagnostic), Department of Radiography and Nursing, Faculty of Health and Wellness Science, Cape Peninsula University of Technology, Cape Town, South Africa.

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
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
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 cracking on palpation 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
epiphyseal plates [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
regimen 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’ diaphysal 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.

References
Butterworth-Heinemann Ltd.
2. Yochum T R & Rowe L J. Essentials of Skeletal Radiography, 2nd
2004, USA: Mosby Inc.
5. Strauss L G. eMedicine: Musculoskeletal. Ewing’s sarcoma. 2007,
28/05/12.
6. Perez CA & Brady L W. Principles and practice of radiation oncology, 2nd
sarcomas: estimation of necrotic fraction during induction chemotherapy
with dynamic contrast-enhanced MR imaging, Radiology. 2003; 228:
271-278.
sarcoma: MR imaging with chemotherapy-induced changes with