Abstract. With advances in imaging technology and increased alertness by clinicians, the reported incidence of primary vertebral osteosarcoma (PVOS) has increased in recent times and, therefore, the importance of its correct diagnosis has repeatedly been emphasized. One such case of PVOS is reported, that presented with insidious clinical, radiological and pathological findings, resulting in a slightly delayed final diagnosis and treatment.

Although osteosarcoma is the most common non-hematological primary malignancy of the bone (1), primary vertebral osteosarcomas (PVOS) account for only 0.8-3% of these tumors (2). An earlier published study reported nine fully documented cases of PVOS over a period of eight decades (3). However, with the advances in imaging technology and the increased awareness of the clinicians, the reported incidence of this perplexing entity has increased in recent times. The latest, large published series from the Mayo Clinic reported an incidence of 4% for PVOS out of the 4,500 cases of osteosarcoma included in the study (1). The tumor presents with a variety of clinical symptoms and radiological findings and, therefore, the importance of differential diagnosis while investigating patients with back pain has repeatedly been emphasized (4). One such case of PVOS is reported, that presented with insidious clinical, radiological and pathological findings, resulting in a slightly delayed final diagnosis and treatment.

Clinical Presentation

The patient was a relatively healthy 79-year-old Caucasian female who presented with gradually worsening right side lower back pain. She had a history of an accidental fall 18 months previously and categorically stated that back pain had commenced after the accident. She also noticed that the pain had gradually worsened in intensity and had started to radiate down her right thigh in the previous few weeks. Walking, bending and performing household chores, as well as coughing and sneezing, all caused aggravation of the pain. However, there were no other symptoms such as tingling, numbness or decreased sensations, neither noticeable weakness in the lower limbs. There was no bowel or bladder dysfunction. Upon deep palpation, tenderness was noticed in the midline of the lumbar spine area. Detailed neuromuscular examination revealed essentially intact functions in the lower limbs with slightly depressed, but bilaterally equal, deep tendon reflexes in both lower limbs. The superficial abdominal and cremaster reflexes were normal with normal sphincter tone.

Radiology

Plain radiographs of the lumbar and sacral spine were obtained in the AP and lateral views with flexion and extension positions. The only positive findings were an old, healed fracture of the L-2 superior end plate and some calcification in the L-3 vertebral body (Figures 1a and 1b). The patient was prescribed pain killers and was requested to come for follow-up 4 weeks later. She complained of further worsening of the pain, despite medication, and was limping significantly in the follow-up visit. The decision to obtain a CT-guided biopsy from the L-3 area was made at the time. Multiple core samples were obtained from the lesion and sent for histological examination. The histopathological examination of the tissue revealed essentially intact functions in the lower limbs with slightly depressed, but bilaterally equal, deep tendon reflexes in both lower limbs. The superficial abdominal and cremaster reflexes were normal with normal sphincter tone.
lumbosacral spine was performed at this time, revealing a destructive exophytic lesion at the L-3 level measuring approximately 35x22 mm. The lesion had a lobulated anterior extension, partially compressing the inferior vena cava and extending between the IVC and aorta (Figure 2). The right transverse process at the L-3 level was fractured. The mass extended to the right side and had displaced the psoas muscle. The differential diagnoses considered radiologically at this time were metastasis, lymphoma, fungal mass, osteomyelitis (tuberculous) or a chondroid neoplasm. Staging CT-imaging studies for her whole body were performed, but did not detect the presence of neoplastic disease at any other location in the body. A subsequent bone scan was performed which revealed a single focus of abnormal tracer activity at the L-3 level, consistent with the imaging findings.

**Intervention**

Considering the discrepancy between the previous CT-guided tissue samples and the nuclear imaging studies, a decision was made to obtain further tissue samples to verify the diagnosis. The patient was taken for a right-side transpedicular open biopsy at the L-3 level using biplanar fluoroscopy guidance, under general anesthesia. The tissue sample was obtained and sent for histology.
Histopathology

Permanent sections of the needle core bone biopsy showed abnormal woven bone infiltrated by a small, round malignant cell neoplasm producing eosinophilic osteoid matrix (Figures 3a, 3b and 3c). The lesional tumor cells were non-reactive to a panel of immunostains for the following markers: low molecular weight cytokeratin, epithelial membrane antigen, S-100 protein, gross cystic disease fluid protein, estrogen receptors, progesterone receptors, leukocyte common antigen, B-cell marker CD 20, T-cell marker CD 3 and vascular marker CD 31. The Congo red stain was negative for amyloid and special tissue stains were negative for fungi and acid-fast bacilli. The sections were examined in consultation with pathologists from the Mayo Clinic, Rochester, MN, USA, who confirmed the diagnosis of grade IV osteoblastic osteosarcoma.

Management

The patient was not considered to be a good candidate for extensive surgical excision and, therefore, was immediately referred to oncologists for chemo-radiation. Since she refused to travel to an out-of-town proton beam facility, radiation therapy was given using external beams. The lumbar spine was irradiated with 36 Gy in 18 fractions, and a further boost of 34 Gy in 17 fractions was delivered to the perivertebral mass at the L-3 level using intensity modulated radiation with the step-and-shoot technique. The therapy was well tolerated with minimum side-effects.

Follow-up

The patient was seen in follow-up, 4 weeks after completion of the radiation treatment, by a team comprising the surgeon, radiation oncologist and medical oncologist. At this time, she reported complete relief from back pain, even with activity. The neurological examination was completely normal. Repeat MR imaging was performed at this time. The scans showed a significant interval reduction in the size of the tumor. The patient was informed about the progress and was also presented with the option of chemotherapy. However, she refused chemotherapy at this point and has been scheduled for regular follow-up.
Figure 3. a, b, c) Permanent sections of the needle core bone biopsy (10x, 45x, 100x) showing abnormal woven bone infiltrated by a small, malignant round cell neoplasm producing an eosinophilic osteoid matrix.


Discussion

Osteosarcomas of the lumbar spine constitute about 1.3% of all osteosarcomas affecting the skeleton (4). The presentation of the tumor in the spine itself has such a wide spectrum of clinical and radiological findings that, combined with the rare incidence, means that it is very commonly missed at first presentation in most patients. Extensive imaging studies, combined with a high degree of clinical suspicion, form the key to an early diagnosis of this potentially treatable tumor. Early detection and accurate diagnosis is important in improving not only the prognosis, but also the quality of the patient’s life (5). The most commonly reported presentation for these patients is backache followed by neurological deficits (6). The symptomatic period between onset and final diagnosis ranges from 2 to 18 months (7). In the present case report, the onset of pain clearly followed an accidental fall in a previously asymptomatic, relatively healthy patient and further added to the confusion regarding diagnosis. Being consistent with the presented case, plain radiographs of these lesions usually demonstrate mineralization of the bone matrix to a varying degree. The typical "ivory vertebra appearance", highly suggestive of the diagnosis, is seen only in 6-7% of the cases (4). The CT study performed in this case at the time of biopsy, however, failed to demonstrate any additional features of the tumor (e.g., mineralized mass, intraspinal extension etc.). Also, spinal canal invasion, which is seen in more than 80% of these tumors, was not detected in the present case. Most recent studies suggest that CT-guided needle biopsy may be the optimal a procedure to obtain a tissue diagnosis, since it minimizes the risk of contamination of the surrounding soft tissue by the tumor cells (7). However, in the current patient’s case, it failed to reveal the malignant nature of the mass. Nevertheless, MRI and bone scan studies definitely indicated a lesion with malignant activity and, therefore, have rightly become an essential part of early investigation for all tumors involving the spine.

An interesting point in the present study was that, despite the aggressive histological category of the tumor and the relative delay (4-6 weeks) between presentation and institution of a definitive therapy, the tumor did not invade the spinal canal and produce irreversible neurological damage. The current trend for the treatment of this aggressive tumor is to use a combined approach with high-dose radiation and multi-drug combination chemotherapy (7, 8). Extensive surgical excision also continues to be a primary option, depending upon the degree of invasion and the general physical condition of the patient (9). In the present case, however, the patient was not considered fit for a major surgical procedure and she categorically refused to receive any aggressive chemotherapy, but yet responded very well to the palliative radiation alone. She continues to attend follow-up and has not demonstrated any signs of disease progression. There have only been a few reports on long-term survivors of spinal osteosarcoma, despite all modalities of treatment. Combination therapy offers the best chance of improved prognosis, but the presented case emphasizes the importance of early diagnosis and even sub-optimal palliative therapy using radiation to achieve tumor-regression and improve the quality of life in elderly patients who might not be considered optimal candidates for aggressive surgical and/or chemotherapeutic intervention.

References


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