The authors describe 10 cases of osteosarcoma of the spine treated between January 1951 and December 2010, and obtained from the Tumour Registry of their hospital. The mean age at presentation was 38.8 years (range: 16-73 years); the mean duration of symptoms was 5.1 months (range: 3 weeks-1 year). Pain was the commonest complaint (9 patients), followed by neurological compromise (6 patients). The thoracic spine and male gender were predominant. Seven patients underwent marginal resection, 3 underwent intralesional resection. All, except one, had adjuvant chemotherapy and radiotherapy, pre- and/or postoperatively. This rare sarcoma has a dismal prognosis: the median survival period was only 2.3 years. The 1-year, 3-year and 5-year survival rates were 80%, 40% and 20%. Astonishingly, marginal resection (7 cases) did not lead to a longer survival than intrallesional resection (3 cases): respectively 30 months and 42 months. Quite logically, local recurrence in 6 patients was linked to a survival of only 36 months, while the other 4 patients survived 52 months. Age below 40 was a positive factor, but not significantly. All patients had a reasonable quality of life with outcomes consistent with the available literature. Recent literature stresses that there is a trend toward improved survival with en bloc resection.

Keywords: osteosarcoma; spine.

INTRODUCTION

Primary osteosarcoma of the spine is rare, accounting for 0.85-3% of all osteosarcomas (1) and for 3.6-14.5% of all primary tumours of the spine (4,10,16). The literature is scarce, due to its low incidence. The optimal treatment strategy is controversial and often guided by the results of various small series and case reports. The treatment of choice is wide excision of the tumour, which has been reported to have a high local control rate (13). Combined chemotherapy and irradiation has been recommended and may be beneficial for some patients. The prognosis of osteosarcoma has remained poor, but in recent years...
aggressive neo-adjuvant and adjuvant chemotherapy have improved the treatment outcome (1,4,15,18, 19,20).

PATIENTS AND METHODS

The hospital Tumour Registry yielded 10 cases between January 1951 and December 2010. All patients were followed-up until death.

The statistical analysis was performed using SPSS (Version 18, Chicago, IL). Kaplan-Meier curves were used for survival analysis. The log-rank (Mantel-Cox) test was used to compare the survival analysis between two patient groups (6 below and 4 above age 40). A p value < 0.05 was seen as statistically significant.

RESULTS

The study population (Table I) comprised 7 male (70%) and 3 female (30%) patients. The mean age at presentation was 38.8 years (range : 16-73 years), the median age 39 years. The mean follow-up period was 2.8 years. All 10 patients were dead at the time of review.

The mean duration of symptoms at presentation was 5.1 months (range : 3 weeks-1 year). Pain was the most common presenting complaint, occurring in 9 patients (90%), followed by neurological compromise in 6 patients (60%). The only patient (case 7) who did not experience any pain presented primarily with cord compression symptoms (paresthesiae which progressed to quadriparesis).

Five of the 9 patients whose initial problem was pain subsequently developed neurological compromise. Three of these had cord compression symptoms (paresthesiae, myelopathy or quadriplegia), while the other two developed a cauda equina syndrome with saddle paresthesiae and incontinence.

Three cervical, 4 thoracic, one lumbar, and 2 sacral localizations were seen. Radiologically, a combination of osteoblastic and osteolytic lesions was noted (Fig. 1-3). All 10 patients had a definite histological diagnosis : primary ‘conventional’ osteogenic osteosarcoma.

All patients were treated surgically. Three patients (30%) had an intralesional resection (including open biopsy), and survived on an average 3 years and 6 months ; 7 patients (70%) had a marginal resection, and survived on average 2 years and 6 months, somewhat paradoxically. Nine patients received pre- and/or postoperative chemotherapy and radiotherapy. Four patients had no local recurrence and survived on an average 4 years and 4 months ; 6 patients had a local recurrence, and survived on an average 3 years and 1 month. The survival analysis of the patients above and below age 40, irrespective of metastases, showed that patients below age 40 survived on average 3 years and 5 months, while those above age 40 survived only 1 year and 9 months. The difference was not statistically significant, probably due to the small number of cases (p = 0.117). Two male patients below age 40 and with thoracic involvement survived more than 5 years. One (case 2) underwent intralesional resection with radiotherapy and adjuvant chemotherapy, and died of persistent disease. The other one (case 3) had a marginal resection without receiving any adjuvant therapy and subsequently had a recurrence at 4 years.

All but one patient died of the disease (persistent disease or lung metastases) with a median survival of 2.3 years. The 1-year, 3-year and 5-year survival rates were 80%, 40% and 20% (Figs. 4 & 5).

DISCUSSION

Most spinal surgeons will see only a few patients with an undiagnosed primary bone tumor of the spine during their entire career (17). Primary osteosarcoma of the spine is extremely rare (1,10,18). The authors found only 10 cases in the hospital Tumor Registry in a 59 year period (1951 through 2010). The mean age at presentation was 38.8, the median age 39, in keeping with other series (10,14). Thus there was a tendency for spinal osteosarcoma to affect the older age group, in sharp contrast with appendicular osteosarcoma (8). There was a male predominance, in contrast to a slight female predominance in other series (10,14). The mean duration of symptoms at presentation was 5.1 months, similar to other reports (2,14).

Clinical picture

The principal modes of presentation for osteosarcoma of the spine are back pain and neurological

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Table I. — Demographics and follow-up data of 10 patients with primary ‘conventional’ osteogenic osteosarcoma

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age / Sex</th>
<th>Site</th>
<th>Duration of symptoms</th>
<th>Clinical presentation symptoms</th>
<th>Pre-operative RT / CT</th>
<th>Surgery</th>
<th>Adjuvant RT / CT</th>
<th>Recurrence / Period after initial management</th>
<th>Survival / Cause of death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20 / F</td>
<td>L4, L5</td>
<td>2 months</td>
<td>Radicular leg pain</td>
<td>N</td>
<td>Marginal resection</td>
<td>Y</td>
<td>N</td>
<td>10 months / Persistent disease</td>
</tr>
<tr>
<td>2</td>
<td>16 / M</td>
<td>T9</td>
<td>1 year</td>
<td>Lower back pain, myelopathy (progressive unsteadiness and paraplegia)</td>
<td>N</td>
<td>Intra-lesional resection</td>
<td>Y</td>
<td>Y / 3 years</td>
<td>5 years and 1 month / Persistent disease</td>
</tr>
<tr>
<td>3</td>
<td>39 / M</td>
<td>T4</td>
<td>9 months</td>
<td>Intercostal root pain</td>
<td>N</td>
<td>Marginal resection</td>
<td>N</td>
<td>Y / 4 years</td>
<td>6 years and 5 months / Persistent disease</td>
</tr>
<tr>
<td>4</td>
<td>20 / M</td>
<td>Sacrum</td>
<td>6 months</td>
<td>Lower back pain, radicular leg pain</td>
<td>N</td>
<td>Intra-lesional resection</td>
<td>Y</td>
<td>N</td>
<td>4 years / Unrelated illness</td>
</tr>
<tr>
<td>5</td>
<td>41 / M</td>
<td>C5, C6</td>
<td>1 year</td>
<td>Neck pain and radicular arm pain</td>
<td>N</td>
<td>Marginal resection</td>
<td>Y</td>
<td>N</td>
<td>2 years and 3 months / Persistent disease</td>
</tr>
<tr>
<td>6</td>
<td>55 / F</td>
<td>C4</td>
<td>1.5 months</td>
<td>Neck pain and quadriplegia</td>
<td>N</td>
<td>Marginal resection</td>
<td>Y</td>
<td>Y / 9 months</td>
<td>12 months / Persistent disease</td>
</tr>
<tr>
<td>7</td>
<td>73 / M</td>
<td>C6–C8, T1</td>
<td>2 months</td>
<td>Upper limb paraesthesia and progressing to quadriparesis</td>
<td>N</td>
<td>Marginal resection</td>
<td>Y</td>
<td>N</td>
<td>2 years and 6 months / Persistent disease</td>
</tr>
<tr>
<td>8</td>
<td>60 / M</td>
<td>T9–T12</td>
<td>5 months</td>
<td>Lower back pain, buttoc paraesthesiae, urinary and faecal incontinence</td>
<td>N</td>
<td>Intra-lesional resection</td>
<td>Y</td>
<td>Y / 4 months</td>
<td>1 year and 5 months / Lung metastases</td>
</tr>
<tr>
<td>9</td>
<td>25 / F</td>
<td>T4–T7</td>
<td>1 month</td>
<td>Lower back pain and loss of sensation from mid-thoracic level down</td>
<td>Y</td>
<td>Marginal resection</td>
<td>Y</td>
<td>Y / 2 years</td>
<td>3 years and 1 month / Lung metastases</td>
</tr>
<tr>
<td>10</td>
<td>39 / M</td>
<td>Sacrum</td>
<td>3 weeks</td>
<td>Lower back pain, radicular leg pain, buttoc paraesthesiae, urinary and faecal incontinence</td>
<td>Y</td>
<td>Marginal resection</td>
<td>Y</td>
<td>Y / 10 months</td>
<td>1 year and 2 months / Persistent disease</td>
</tr>
</tbody>
</table>

NB : M = Male, F = Female, C = Cervical, T = Thoracic, L = Lumbar, RT = Radiotherapy, CT = Chemotherapy, Y = Yes, N = No.
deficit (1,2,5,7,10,14). The authors noted a similar trend: a majority (90%) experienced non-mechanical back or radicular pain, followed in 60% by significant neurological compromise (either cord or cauda equina compression). Two patients with initial back pain developed an early neurological deficit which progressed to quadriplegia in 1.5 and 2 months. Patients presenting with non-mechanical back pain along with rapid neurological deterioration should raise the spectre of a primary spinal malignancy.

Osteosarcoma has been reported to occur at all levels of the spine, most frequently at the sacral level, followed by the lumbar, thoracic and cervical levels (5). In the current series, the most common site was the thoracic spine, followed by the cervical, sacral and lumbar spine.

Plain radiographs (14) and CT-scans display a mixture of osteoblastic and osteolytic areas. However, purely osteolytic lesions (moth-eaten pattern) have been reported in previous series at a rate of 13.7% to 35% (3,5,6,10). These can be misdiagnosed as benign osteoblastomas (12). Chondrosarcoma and sclerotic metastases can also mimic the radiological appearance of pure sclerotic osteosarcoma (ivory, dense cloud-like appearance). Radiological features favoring the diagnosis of primary malignant tumour include: soft tissue mass, periosteal reaction, long lesion (> 6 cm), expansion of bone, and solitary lesion (21). These tumours commonly invade the spinal canal as a soft-tissue mass, which makes cross-sectional imaging like CT-scan and magnetic resonance imaging (MRI) essential for staging and treatment.
Improving the prognosis

The prognosis of primary osteosarcoma of the spine is dismal. Shives et al (14) report a one year survival rate of 26.9% in 26 patients treated between 1909 and 1980. Other authors (1,15) note a median survival rate of 6 to 10 months. A combination of complete resection and intensive chemotherapy is often recommended for the treatment of primary spinal tumors (9,10,14,15): the overall survival is better when compared to patients without this modality. Ozaki et al (10) report a better outcome after aggressive interdisciplinary treatment: an 86% survival rate of > 1 year and a 13.6% survival rate of > 6 years. In the current series, 9 out of 10 patients were treated with a combination of surgery and pre- and/or postoperative chemo- and radiotherapy. Only two patients received neoadjuvant chemo/radiotherapy, marginal resection and postoperative chemo/radiotherapy. They had a dismal survival of one year and 2 months, and 3 years and one month respectively. Eighty percent of patients survived > 1 year with a median survival of 2.3 years. The 3-year and 5-year survival rates were 40% and 20%. These results are comparable to those of other authors (9,10,12). Two patients in the current series had a greater than 5 year survival. Both were male patients below age 40 with osteosarcomas involving the thoracic spine. Only one

planning. A CT-scan is also useful for the diagnosis of soft tissue involvement and lung metastases (11). CT-guided needle biopsy may be optimal to prevent contamination with tumour tissue (7). MRI is superior to CT-scan in assessing intra-osseous tumour spread and neural compression or invasion (5).

There are several different histological variants of osteosarcoma, based on the predominant tissue (osteoblastic, chondroblastic or fibroblastic); “conventional” osteoblastic osteosarcoma is the commonest. In the current series all patients had a histologically confirmed ‘conventional’ osteoblastic osteosarcoma.

Surgery

The aim of spinal surgery is usually palliative rather than curative, due to the aggressive nature of osteosarcoma with soft tissue extension and involvement of multiple levels of the spine. Palliative management should focus on pain control and management of acute cord or cauda equina compression through surgical decompression and stabilization. Surgery may produce a transient improvement of neurological signs and symptoms, but curative treatment is rarely feasible or possible. Indeed, local recurrence occurred in 6 out of 10 patients (60%), on an average 1.8 years after the initial management. Four of the 6 patients with local recurrence were initially managed with marginal resection, and the two others with intralesional resection.

Fig. 4. — Overall survival of 10 patients with primary spinal osteosarcoma.

Fig. 5. — Survival analysis of patients above and below age 40 with primary spinal osteosarcoma.
therapy may of be benefit selected patients. In one of the two patients who received marginal resection, survival was only 1 year. In the other patient, who received wide resection, survival was only 1 year 2 months in spite of absence of co-morbidities. Patients should be counseled in detail about the likelihood of persistent primary disease, local recurrence and metastatic lesions regardless of treatment modality.

Nine of ten patients had persistent primary disease at the time of death regardless of the evolution of treatment modalities in the past six decades. This study has some weaknesses, essentially its retrospective character and the small number of cases. However, no patients were lost to follow-up, and all the case records were reviewed thoroughly.

REFERENCES