Case Report: Localized Ewing’s sarcoma of the scapula in an adult [version 1; peer review: awaiting peer review]

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Abstract
Background: Ewing's sarcoma (ES) of the scapula is a rare entity. It is often discovered late at the metastatic stage of the disease because of its deep location. This neoplasm is common in children and adolescents. We present the first reported case of a localized Ewing's sarcoma of the scapula in an adult over 40 years-old.

Case presentation: A 48-year-old man presented with left shoulder pain evolving for one year. Physical examination showed a painful, ill-defined swelling of the left shoulder measuring 5 x 3 cm. Magnetic resonance imaging (MRI) was performed showing a mass of the left scapula invading the soft tissues of the shoulder suggestive of a sarcoma. The patient underwent a surgical biopsy finding an ES of the scapula with no secondary localization on the computerized tomography (CT) scan nor on the bone scintigraphy. Neo-adjuvant multiagent chemotherapy was started obtaining a total response. Therefore, the patient underwent a total left scapulectomy. Histopathological examination confirmed the diagnosis of ES with a complete response to chemotherapy. Adjuvant chemotherapy was then indicated. After 3 years of follow-up, no local or distant recurrence was found.

Discussion: ES is a high-grade aggressive lesion that most commonly originates in bone. The ES may affect any bone but is frequent in femur, tibia and ilium, the tumors arising from the scapula comprise fewer than 4% of all ES. No cases of localized Ewing's sarcoma in adults have been reported to our knowledge. The diagnosis is confirmed by immunohistochemical examination and cytogenic. A multimodal treatment approach including a combination of chemotherapy, surgery, and radiation can modestly improve local tumor outcomes. Metastatic tumors still have poor diagnosis.

Conclusions: ES occurs rarely in adults greater than 20 years-old, and tumors localized in the scapula are even rarer. Histopathology differentiates it from other primary bone/soft tissues tumors.

Keywords
Ewing's sarcoma, Scapula, Tumors
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**Introduction**

Ewing’s sarcoma (ES) is an aggressive primary osseous neoplasm and those arising from the scapula comprise fewer than 4% of all ES. ES of the scapula are often asymptomatic and grow quite large before being diagnosed due to their deep location, thus these tumors are discovered late and usually at the metastatic stage of the disease. This neoplasm is the second most common malignant bone tumor of children and young adults, but its incidence declines rapidly as age increases beyond 20 years and is extremely rare in adults over 40 years of age. Our literature review showed only two reported cases of Ewing’s sarcoma of the scapula in adults over 40 years old which were discovered at the metastatic stage (cases with insufficient data information were excluded). Here, we present the first case reported to our knowledge of a localized ES of the scapula in an adult over 40 years of age.

This case report has been reported in line with the SCARE Criteria.

**Case presentation**

A 48-year-old male Caucasian police officer presented with a painful swelling of the left shoulder evolving for one year. The patient had no medical or surgical history nor allergies. There was no history of systemic symptoms such as fever, anorexia, or recent subjective weight loss. The increase in size of the swollen shoulder brought him to seek medical attention without taking any medication so far. Physical examination showed a hard, painful, and ill-defined bony mass of the left scapular region measuring 5 × 3 cm without adjacent cutaneous lesions. The mobility of the upper limb was preserved. Cardiovascular, respiratory, and abdominal examinations were unremarkable. An X-ray of the left shoulder showed a bony mass with osteolytic lesions suggestive of a primary bone tumor. Magnetic resonance imaging (MRI) was performed showing a necrotic mass invading the scapula and the soft tissues of the left shoulder suggesting a sarcoma (Figure 1). A full body computerized tomography (CT) scan and bone scintigraphy didn’t show any other localization of

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**Figure 1.** MRI of the scapula.

**Figure 2.** Scapula resection piece.
the tumor. The patient underwent a surgical biopsy of the mass. The intervention was performed by a group of senior orthopedic surgeons in an orthopedic surgery department of a university hospital in Tunisia (Figure 2). Histopathological examination found a malignant tumoral proliferation with large areas of necrosis. The tumor was made of small round cells which have a poorly distributed cytoplasm with ill-defined borders and a vacuolated aspect rich in periodic acid Shiff (PAS) positive glycogen. Immunohistochemically these cells were cluster designation (CD20) negative, Desmine and Myogenine negative, cytoKeratine (CK) and CD56 negative, and were positive for CD99 with a heavy membranous marking. The immunohistochemical results also demonstrated a diffuse nuclear positivity for the NKX2.2. The Ki67 proliferative index was 70% (Figures 3, 4, 5). Thus, the diagnosis of Ewing’s sarcoma of the left scapula was confirmed. After a multidisciplinary meeting, the patient had six cycles of neo-adjuvant vincristine, Ifosfamide, doxorubicin, etoposide (VIDE) chemotherapy. This involved Vincristine 1.5 mg/m² (max 2 mg) intravenous IV Day 1 Doxorubicin

Figure 3. Malignant tumor proliferation, of solid architecture, densely cellular with round cells.

Figure 4. Diffuse nuclear positivity for the NKX2.2.
20 mg/m² IV Days 1, 2 and 3 of cycle Etoposide 150 mg/m² IV Days 1, 2 and 3 of cycle Ifosfamide 3 g/m² IV Days 1, 2 and 3 of cycle. The protocol is repeated every 21 days 06 cycles.

An intermediate MRI showed a total response to the treatment. He then underwent an en-bloc total left scapulectomy carrying the scapula, the infraspinatus muscle, the supraspinatus muscle, and the upper part of the deltoid muscle. No postoperative problems were noticed. Histopathological examination confirmed the diagnosis of ES, with no residual viable tumor cells Grade IV according to Huvos’ criteria and wide resection margins. The patient continued the protocol with an adjuvant VIDE chemotherapy. After 3 years of clinical and radiological follow-up, an MRI was performed showing the absence of local recurrency. Clinically, the patient has a preserved mobility of the upper limb, arm flexion is possible, and he is capable of doing the hand to mouth movement.

Discussion

Ewing’s Sarcoma (ES) was first described by James Ewing in 1925.6 ES is a primary osseous neoplasm. It is also part of the Ewing sarcoma tumor family, which includes primitive neuroectodermal tumor, Ewing soft tissue sarcoma, and Askins tumor.6,7 It accounts for 8% of all malignancies and 2% of all primary bone tumors.2,8 ES is a high-grade aggressive lesion, most commonly of bony origin, with large soft-tissue masses and frequent metastases. 15–30% of patients have distant metastases at diagnosis.8,9 The sites of metastasis are mainly lung (85%), bone (69%), pleura (46%), but also lymph nodes (40%) and central nervous system (12%). 4110,11; ES is the second most common malignant bone tumor in children and young adults. It occurs primarily in the first decade of life (mean age 14 years) and has a slight preference for males. Incidence declines rapidly with increasing age after age 20.4 The average age at diagnosis he is 15 years.2,12 ES is diagnosed in Caucasian Caucasians <25 years of age with an incidence of 0.3/100,000 per year, but is very rare in African and Asian populations.2,13 The ES may affect any bone but is frequent in femur, tibia and ilium and the tumors arising from the scapula comprise fewer than 4% of all ES.1,3 Patients with ES of the scapula are slightly older at time of diagnosis, since these tumors are often asymptomatic and grow quite large before being diagnosed.1,2 Thus, these tumors are usually discovered late, in the metastatic stage of the disease. No cases of localized ES in adults have been reported to our knowledge. The reported cases of metastatic ES of the scapula in adults over 40 years of age are summarized in Table 1.

Clinically, ES commonly presents as a dull to severe persistent shoulder pain that gradually increases in size. Although the etiology of the ES remains unknown, it has been confirmed that majority of cases had a cytogenetic translocation consisting of Rearrangements of the EWS gene and a member of the ETS gene family (FLI1) on chromosome 22q12. These translocations define the Ewing sarcoma tumor family (ESFT) and provide valuable tools for accurate and unambiguous diagnosis.5,14 Radiologically, ES presents as an ill-defined osteolytic lesion, Permeable or worm-eaten bone destruction, often associated with an onion slit-type multi-layered periosteal reaction. On the MRI, the lesion is classically

Figure 5. Cluster of designation (CD99) positivity with a heavy membranous marking.
described as a prominent mass that contains areas of necrosis or hemorrhage. Histopathologically, typical ES is made of uniform small round blue cells with a clear cytoplasm and distant cellular borders, has a uniform small oval blue nucleus. The principal differential diagnosis with ES are small cell osteosarcoma, mesenchymal chondrosarcoma, lymphoma and metastatic neuroblastoma. The diagnosis is confirmed by immunohistochemical examination showing a diffuse strong cytoplasmic membrane positivity for CD99 combined with NKX2.2 positivity and cytogenic examination showing the t (11;22) (q24;q12) translocation. Neither the NKX2.2 nor CD99 alone are entirely specific for the Ewing’s family tumors, but when combined the diagnostic specificity is high. NKX2.2 is positive in 91.5% of the tumors, and CD99 is positive in 99%.

Before the advent of modern chemotherapy, fewer than 10% of her ES patients survived more than 5 years after diagnosis when treated with surgery or radiotherapy alone, but it increases to 60–70% in localized tumors when neo-adjuvant and adjuvant chemotherapy are used in conjunction with surgery/radiotherapy. Currently, the treatment of the Ewing’s sarcoma Coordinated through cooperative groups. Although multimodality treatment approaches involving a combination of chemotherapy, surgery, and radiation have resulted in modest improvements in localized tumor outcomes. Prognosis remains poor for patients with metastatic disease.

The most common surgical approach of the malignant tumors of the scapula was the forequarter amputation until 1970, after that the Tikhoff-Linberg procedure was used. The most used protocol for ES is an intense multiagent neoadjuvant chemotherapy, followed by en bloc excision of the tumor mass and radiotherapy postoperatively if doubt of tumor residue which was achieved with our patient. Adjuvant chemotherapy for consolidation is decided according to the response of the tumor to chemotherapy (Huvos tumor necrosis grading system). Prognostic factors may define subgroups requiring different treatment intensities, the most common are age of the patient, tumor size and location, metastatic pattern, and histologic response to chemotherapy. For the patients with ES of the scapula, those who have metastatic disease, marginal resection, or a chemotherapy response of <80% (Grade I and II of Huvos’ criteria) have the worst prognosis.

### Conclusion
The ES is common in children and adolescent’s long bones and pelvis, but ES of the scapula in adults is a very rare entity. It is often discovered after a diffuse local progression and distant metastases. Diagnosis is confirmed histologically and immunohistochemically. Neo-adjuvant chemotherapy combined with surgery is the most common treatment for localized tumors.

### Consent
Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

### Data availability
**Underlying data**

Data are available under the terms of the Creative Commons Attribution 4.0 International license (CC-BY 4.0).

### References

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Table 1. Reported cases of Ewing’s sarcoma of the scapula in adults over 40 years old.

<table>
<thead>
<tr>
<th>Authors</th>
<th>Gender</th>
<th>Age</th>
<th>Tumor site</th>
<th>Metastases</th>
<th>Treatment</th>
<th>Follow-up (Months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mavrogenis et al. (2009)</td>
<td>M</td>
<td>57</td>
<td>Scapula</td>
<td>Lung</td>
<td>Scapulectomy CT</td>
<td>12</td>
</tr>
<tr>
<td>Malik et al. (2020)</td>
<td>M</td>
<td>51</td>
<td>Scapula</td>
<td>Multimetastases</td>
<td>DOD</td>
<td></td>
</tr>
</tbody>
</table>

M: male, CT: chemotherapy, DOD: died of disease.
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