Head and neck soft tissue sarcomas: prognostic factors and outcome in a series of patients treated at a single institution

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Background: Head and neck soft tissue sarcomas (STS) represent a rare disease.

Patients and methods: One hundred and sixty-seven patients underwent surgery at our institution with an eradicating intent between 1990 and 2010. Local recurrence (LR), distant metastasis (DM) and disease-specific mortality (DSM) incidence were studied along with clinicopathological prognostic factors.

Results: Ten-year crude cumulative incidence (CCI) of LR, DM and DSM were 19%, 11% and 26%, respectively (median follow-up 66 months). Independent prognostic factors for DSM were tumor size ($P < 0.001$) and grade ($P = 0.032$), while surgical margins obtained a border-line significance (0.070); LR was affected by the tumor size ($P = 0.001$), while DM only by grade ($P = 0.047$). The median survival after LR and DM were 14 months and 7 months, respectively. Tumors sited in the paranasal sinus and supraclavicular region had the worst survival.

Conclusions: Head and neck represent a very critical anatomical site for STS. Achievement of local disease control appears to be crucial, since even LR could be a life-threatening event.

Key words: head and neck, prognosis, sarcoma, soft tissue sarcoma, surgery

introduction

Head and neck soft tissue sarcomas (HNSTS) are very rare. In fact, soft tissue sarcomas (STS) account for <1% of all malignant tumors in adults [1], and only 5%–10% of them are localized in the head and neck region [2–4].

The available published series are consequently very few. Moreover, they are often collected across many decades. Entities like bone sarcomas, primitive neuroectodermal tumors (PNETs), pediatric-type rhabdomyosarcoma and desmoid-type fibromatosis are usually included, despite their different biological behavior.

The present study is a retrospective analysis of HNSTS treated at Fondazione IRCCS Istituto Nazionale dei Tumori Milan, Italy (INT) over the last 20 years. The purpose of the study is to analyze prognostic factors and to explore oncologic outcomes in a homogeneous group of adult-type HNSTS treated at a single referral institution.

materials and methods

Between January 1990 and January 2010, overall 3715 consecutive patients affected by localized STS were surgically treated at INT. Among these patients, 206 (6%) had HNSTS. We excluded patients with metastatic spread at referral and patients with the following histologies: PNET/Ewing's family tumor, pediatric-type (alveolar and embryonal) rhabdomyosarcoma and desmoid-type aggressive fibromatosis. Bone sarcomas were a priori excluded.

Eventually, patient series included 167 cases.

Clinical data were prospectively collected in our institutional database and retrospectively reviewed.

The anatomical site was classified as follows: scalp and face, paranasal sinus, neck without supraclavicular and supraclavicular fossa.

Histological diagnosis was reviewed in all cases by two experienced pathologists at our institution.

As far as histological grading is concerned, the French Federation of Cancer Centers (FNCLCC) grading system [5] was applied to the untreated primary tumors. In the case of recurrent tumor, the grading was carried out on the slides from the primary untreated tumor seen in consultation.

With regard to treatment, every patient underwent surgery at INT with an eradicating intent. Surgical specimens were routinely examined in the presence of the operating surgeon.

Surgical excisions were considered as macroscopically complete in the absence of gross residual disease. Consistently with other reports [3,6–10],
all macroscopically complete resections were classified according to the closest surgical margin, which was microscopically categorized as positive (tumor within 1 mm from the inked surface) or negative (absence of tumor within 1 mm from the inked surface).

Resections were categorized as intralesional if a macroscopic residual disease was left behind.

For sarcomas of paranasal sinus, due to the peculiar anatomy, margin classification was adapted according to the presence of reliable anatomical barrier to neoplastic diffusion (i.e. dura mater and bone). Negative margins were therefore defined as no infiltration or partial infiltration of the barrier removed around the tumor; positive margins if the barrier was infiltrated throughout its entire thickness; and intralesional as described above.

The indication to radiotherapy (RT) was considered in the case of positive surgical margins not amenable to further surgical re-excision and/or high grade sarcoma [11].

External beam radiation was delivered by IMRT (intensity modulated RT) or VMAT (volumetric modulated arc therapy) with 6 MV photons. The delivered dose was at least 60–62 Gy in the case of negative margins and at least 64–66 Gy in the case of positive or close margins, in fractions of 2 Gy each.

Table 1. Main tumor and treatment characteristics according to site and presentation

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<th>Neck</th>
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<td>≤5</td>
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<td>11 (9%)</td>
<td>5 (11%)</td>
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<td>3 (7%)</td>
<td>12 (31%)</td>
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<td>4 (9%)</td>
<td>19 (11%)</td>
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<td>2 (5%)</td>
<td>6 (16%)</td>
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<td>3 (7%)</td>
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<td>15 (88%)</td>
<td>24 (53%)</td>
<td>9 (24%)</td>
<td>50 (40%)</td>
<td>11 (25%)</td>
<td>61 (37%)</td>
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<tr>
<td>Epidermal graft</td>
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<td>0</td>
<td>3 (7%)</td>
<td>0</td>
<td>6 (5%)</td>
<td>3 (7%)</td>
<td>9 (5%)</td>
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<tr>
<td>Local flap</td>
<td>46 (69%)</td>
<td>0</td>
<td>11 (24%)</td>
<td>4 (10%)</td>
<td>39 (32%)</td>
<td>22 (50%)</td>
<td>61 (37%)</td>
</tr>
<tr>
<td>Pedicled flap</td>
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<td>1 (6%)</td>
<td>7 (16%)</td>
<td>15 (40%)</td>
<td>18 (15%)</td>
<td>6 (14%)</td>
<td>24 (14%)</td>
</tr>
<tr>
<td>Free flap</td>
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<td>1 (6%)</td>
<td>0</td>
<td>10 (26%)</td>
<td>10 (8%)</td>
<td>2 (4%)</td>
<td>12 (7%)</td>
</tr>
</tbody>
</table>

*aOther includes fibrous solitary tumors, pleomorphic rhabdomyosarcomas, extraskeletal chondrosarcoma, histiocytic sarcoma, malignant epithelioid angiomylipoma, sclerosing epithelioid.

SC, supraclavicular; PS, paranasal sinus; NA, not available; n.a., not applicable. The percentages are referred to the total number of the columns.
Neoadjuvant/adjuvant chemotherapy (CT) was offered to those patients carrying a lesion with a high risk of recurrence (i.e. high grade, deep location and size >5 cm). An anthracycline-based CT was prevalently used. In most cases, anthracycline was associated with ifosfamide.

After surgery all patients underwent surveillance at our institution as follows: re-evaluation every 4 months in the first 2 years, every 6 months in the following 3 years and every year in the following 5 years. At every time point at least clinical examination, ultrasonography or MRI for local staging and chest X-ray or chest CT scan were carried out. Further examination to assess metastatic spread was required depending on clinical suspicion.

**statistical methods**

The prognostic analyses mainly focused on investigating the prognostic effect of the anatomical site, classified as: (i) scalp and face, (ii) neck, (iii) supraclavicular, (iv) paranasal sinus, on the following end points: disease-specific mortality (DSM), local recurrence (LR), and distant metastasis (DM). Concomitant local relapses and distant metastases were included in the computations as distant metastases. The analyses were carried out in a competing risks framework; in the analysis of DSM, deaths due to conditions unrelated with sarcoma were regarded as competing events. For LR (DM) analysis, deaths without evidence of disease and DM (LR), whichever occurred first, were regarded as competing events. Event times were computed from the date of surgery at our institution, or censored at the date of last follow-up assessment for event-free patients.

Additional putative prognostic factors were: presentation (primary, recurrence), depth (superficial, deep), surgical margins (negative, positive, intralesional), tumor size, histology (dermatofibrosarcoma protuberans (DFSP), malignant peripheral nerve sheath tumor (MPNST), leiomyosarcoma, malignant fibrous histiocytoma (MFH)/unclassified pleomorphic sarcoma (UPS), others), and FNCLCC grade (I, II, III).

Univariable analyses were carried out by estimating the LR, DM and DSM crude cumulative incidence (CCI) curves. Between-curves comparison was carried out by using the Gray test [12]. Multivariable analyses were based on cause-specific hazards and were therefore carried out using Cox regression models [13]. To control for overfitting due to the model high dimensionality when compared with the low number of events, penalized maximum likelihood estimation methods [14] were applied as implemented in the R package penalized. Patient age and tumour size were modeled as continuous variables by using four-knot restricted cubic splines [15] to obtain flexible fits, by allowing their prognostic effect not to be the same in every part of the range, whereas the other categorical prognostic factors were modeled by using dummy variables.

Additional analyses were based on Kaplan–Meier method [16] for overall survival curves estimation; the time was computed from the date of surgery to the date of death due to any cause, or censored at the date of last follow-up assessment for alive patients.

![Figure 1](image-url). Local relapse (A), distant metastasis (DM) (B) and disease-specific mortality (DSM) (C) crude cumulative incidence (CCI) curves according to anatomical site of tumor.
Standard descriptive statistics were calculated for categorical data (i.e. frequency and percentage) and continuous data (i.e. median and interquartile [IQ] range), as listed in the tables.

The association between the anatomic site and other prognostic factors was tested using the Kruskal–Wallis test for continuous prognostic factors (i.e. patient’s age, tumor size) and the exact Chi-square test for categorical ones.

We considered as significant two-sided *P* values below the 5% conventional threshold. All statistical analyses were carried out using the SAS (SAS Institute Inc., Cary, NC) and the R software [17].

**results**

Clinicopathological data are summarized in Table 1.

As of September 2010, the median duration of follow-up was 66 months (IQ range 37–120 months).

**local recurrence**

Thirty-five events were recorded after surgery at our institution: 26 among 119 primary cases (22%) and 9 among 44 recurrent cases (20%). In 27 cases, LR was unique, while 8 patients had more than one relapse. For patients who experienced it the median time to first LR was 16 months.

Twenty-four patients presented only local relapse of the disease, while in 11 cases it was associated with metastases.

Twenty-five relapsed patients subsequently died of disease (71%). The median post-relapse survival was 14 months (range 3–24).

Ten-year CCI of LR was 19% (13.2%–27.4%) in the whole series.

According to univariable analysis (supplementary Table S1, available at *Annals of Oncology* online), all investigated prognostic factors but presentation were associated with LR risk. In fact 10-year CCI of LR was 18.8% in primary cases versus 20.6% in recurrent ones (Gray test *P* = 0.759).

The CCI of LR according to the anatomical site, histology and grade is depicted in Figures 1–3. Paranasal sinus tumors showed a higher risk of LR (10–year CCI: 41.6%), as well as MPNST (34.3%) and leiomyosarcoma (28.5%) among the histotypes. Only two LRs were recorded among DFSP (4.5%). As expected, in the case of negative surgical margins the LR incidence was much lower (13.6%) than in the case of positive margins (28.5%) and intralesional margins (50.0%) (*P* = 0.010).

At multivariable analysis, the only independent prognostic factor was tumor size (*P* = 0.001, Table 2).

Figure 2. Local relapse (A), distant metastasis (DM) (B) and disease-specific mortality (DSM) (C) crude cumulative incidence (CCI) curves according to histology.
Ten-year incidence estimate of local relapse was 27%, when excluding DFSP patients.

distant metastases
Nineteen patients experienced DM: 15 among 119 primary tumors (13%) and 4 among 44 recurrent tumors (9%).
For patients who had it time to metastatic spread varied from 2 to 41 months, with a median value of 9 months.
Sites of metastatic spread were: lung (10 out of 19, 59%), lymph nodes (5, 29%), bone (4, 23%), brain and soft tissue (2 each, 12%), liver (1, 6%). Five patients had multiple metastatic sites. Sixteen deaths (84%) were recorded in this group. The median post-metastasis survival was 7 months (range: 2–42 months).
Ten–year CCI of DM was 10.7% (95% CI 6.7%–17.0%).
At univariable analysis (supplementary Table S1, available at Annals of Oncology online), all the investigated prognostic factors but presentation and surgical margins were associated with DM risk; in primary cases 10–year DM CCI estimate was 10.8%, while in recurrent cases it was 10.4% (Gray test, $P = 0.867$).
CCI of DM according to anatomical site, histology and grade are shown in Figures 1–3.
In case of tumor sited in paranasal sinus, incidence of DM was far worse (10–year CCI: 26.3%). According to histotypes, leiomyosarcoma (33.3%) and MPNST (26.1%) showed the strongest trend to metastatic spread, while none of DFSP metastasized.
At multivariable analysis (Table 2), the only independent prognostic factor was FNCLCC grade ($P = 0.047$); tumor depth obtained a borderline $P$ (0.069).
Ten-year incidence estimate of DM was 16%, when excluding DFSP patients.

disease-specific mortality
Overall 43 deaths were recorded. Thirty-six were disease-related events: 20 consequent to local tumor progression (16 because of local relapse and 4 as persistence of disease) and 16 to
metastatic spread. Cause-specific deaths were 24 among 123 primary tumors (19%) and 12 among 44 recurrent cases (27%).

In the whole series, overall survival and CCI of DSM at 10 years (95% confidence interval) were 66% (57.2%–76%) and 26% (19.5%–34.7%).

In univariable analysis (supplementary Table S1, available at *Annals of Oncology* online), all the putative prognostic factors but presentation were statistically significant. In particular, in Figures 1–3, DSM CCI curves are depicted according to the anatomical site, histology and grade. As for the anatomical site, scalp and face (10–year cumulative incidence: 4.7%) and neck (10–year cumulative incidence: 4.7%) fared much better than supraclavicular (32.1%) and paranasal sinus (54.3%). According to the histotype, the worst mortality was observed for leiomyosarcoma (62.6%) and MPNST (48.6%), while only one patient affected by DFSP died because of disease (1.9%). Eventually, adequate surgical treatment was significantly linked to a lower mortality rate (18.9% in case of negative margins versus 39.2% and 66.7% in the case of positive and intralesional margins, respectively).

At multivariable analysis (Table 2), tumor size and FNCLCC grade were confirmed as independent prognostic factors ($P < 0.001$ and $P = 0.032$, respectively). Anatomical site did not reach significance ($P = 0.362$), while surgical margins obtained a borderline $P (0.070)$.

Ten-year incidence estimate of DSM was 39%, when excluding DFSP patients.

### anatomical site

The anatomical site was significantly associated with size ($P < 0.0001$), depth ($P < 0.0001$), histotype ($P < 0.0001$) and grade ($P < 0.0001$). In detail, in supraclavicular and paranasal sinus there was a higher percentage of larger, deeply located, high-grade sarcomas. Moreover, leiomyosarcomas were the most frequent histotype in paranasal sinus, as well as MPNST in supraclavicular. Conversely, DFSP were mainly located in scalp and face.

As already cited, sarcomas sited in paranasal sinus showed the worst survival estimates, followed by those sited in the supraclavicular region.

### peculiar cases

In five cases (3%), patients had undergone previous RT in pediatric age for other disease (mainly lymphoma), so that radioinduced etiology could be hypothesized. Histology was NOS sarcoma in four cases and leiomyosarcoma in one. All these sarcomas were high-risk (high-grade and mean diameter 9.2 cm) and they had overall very aggressive behavior: three recurred locally and two metastasized. Patients’ prognosis was consequently particularly severe: two died because of local progression, two had incurable metastatic disease and only one is still disease-free.

Three patients (2%) were affected by Von Recklinghausen disease; two of them had MPNST (2 out of 17 MPNST, 17%) and one presented a DFSP. Among them, one patient affected by MPNST presented LR despite postoperative RT/CT treatment and died because of local progression.

### discussion

In this series of 167 patients affected by HNSTS treated at our institution over a 20-year time span 10-year incidence estimates of local relapse, DM and DSM were 19%, 11% and
## Table 3. Comparison of major and most recent series of head and neck soft tissue sarcomas

<table>
<thead>
<tr>
<th>Reference</th>
<th>No of centers</th>
<th>Pts</th>
<th>Study period</th>
<th>Peculiar histotypes included</th>
<th>% pts surgically treated</th>
<th>% Negative margins</th>
<th>Median FU (mos)</th>
<th>Primary at referral</th>
<th>Metastatic at referral</th>
<th>5y LRFS</th>
<th>5y DMFS</th>
<th>5y DSS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weber et al. [18]</td>
<td>1 (a)</td>
<td>188</td>
<td>1960–1982</td>
<td>Desmoid</td>
<td>70%</td>
<td>62%</td>
<td>29</td>
<td>N.S.</td>
<td>17 (9%)</td>
<td>N.S.</td>
<td>N.S.</td>
<td>N.S.</td>
</tr>
<tr>
<td>Farhood et al. [19]</td>
<td>1 (b)</td>
<td>176</td>
<td>1950–1985</td>
<td>–</td>
<td>90%</td>
<td>32%</td>
<td>N.S.</td>
<td>176 (100%)</td>
<td>0</td>
<td>N.S.</td>
<td>N.S.</td>
<td>OS: 55%</td>
</tr>
<tr>
<td>Wanebo et al. [20]</td>
<td>33 (c)</td>
<td>214</td>
<td>1982–1990</td>
<td>Bone sarcomas; Desmoid;</td>
<td>84%</td>
<td>76%</td>
<td>N.S.</td>
<td>189 (88%)</td>
<td>0</td>
<td>(186 pts)</td>
<td>DFS: 57%</td>
<td>(203 pts)</td>
</tr>
<tr>
<td>Eeles et al. [21]</td>
<td>1 (d)</td>
<td>103</td>
<td>1944–1988</td>
<td>–</td>
<td>77%</td>
<td>14% (n)</td>
<td>50</td>
<td>N.S.</td>
<td>2 (2%)</td>
<td>47%</td>
<td>68%</td>
<td>OS: 70%</td>
</tr>
<tr>
<td>Kowalski et al. [20]</td>
<td>1 (e)</td>
<td>128</td>
<td>1953–1985</td>
<td>–</td>
<td>73%</td>
<td>72%</td>
<td>N.S.</td>
<td>66 (52%)</td>
<td>16 (12%)</td>
<td>DFS: 35%</td>
<td>OS: 48%</td>
<td></td>
</tr>
<tr>
<td>Le et al. [23]</td>
<td>1 (f)</td>
<td>65</td>
<td>1961–1993</td>
<td>Desmoid</td>
<td>83%</td>
<td>35%</td>
<td>64</td>
<td>65 (100%)</td>
<td>0</td>
<td>66%</td>
<td>86%</td>
<td>60%</td>
</tr>
<tr>
<td>Dudhat et al. [24]</td>
<td>1 (g)</td>
<td>72</td>
<td>1981–1995</td>
<td>–</td>
<td>100%</td>
<td>75%</td>
<td>38</td>
<td>60 (83%)</td>
<td>0</td>
<td>DFS: 45%</td>
<td>OS: 60%</td>
<td></td>
</tr>
<tr>
<td>Bentz et al. [2]</td>
<td>1 (h)</td>
<td>111</td>
<td>1973–1999</td>
<td>Desmoid (excluded from DSS)</td>
<td>94%</td>
<td>52%</td>
<td>51</td>
<td>62 (56%)</td>
<td>37 (33%)</td>
<td>DFS: 55%</td>
<td>(89 pts)</td>
<td>52%</td>
</tr>
<tr>
<td>De Bree et al. [25]</td>
<td>1 (i)</td>
<td>38</td>
<td>1983–2004</td>
<td>Ewing’s sarcoma</td>
<td>87%</td>
<td>18%</td>
<td>73</td>
<td>N.S.</td>
<td>2 (5%)</td>
<td>DFS: 52%</td>
<td>OS: 81%</td>
<td></td>
</tr>
<tr>
<td>Mucke et al. [26]</td>
<td>2 (l)</td>
<td>74</td>
<td>1999–2006 and 2000–2008</td>
<td>Bone sarcomas</td>
<td>100%</td>
<td>92%</td>
<td>43</td>
<td>74 (100%)</td>
<td>0</td>
<td>N.S.</td>
<td>N.S.</td>
<td>OS: 61%</td>
</tr>
<tr>
<td>Van Damme et al. [27]</td>
<td>1 (m)</td>
<td>42</td>
<td>1988–2008</td>
<td>–</td>
<td>93%</td>
<td>26%</td>
<td>54</td>
<td>N.S.</td>
<td>4 (10%)</td>
<td>DFS: 47%</td>
<td>OS: 72%</td>
<td></td>
</tr>
<tr>
<td>Current series</td>
<td>INT</td>
<td>167</td>
<td>1990–2010</td>
<td>–</td>
<td>100%</td>
<td>68%</td>
<td>66</td>
<td>123 (74%)</td>
<td>0</td>
<td>LRI: 17%</td>
<td>DMI: 11%</td>
<td>CSM: 26%</td>
</tr>
</tbody>
</table>

(a) University of Texas, MD Anderson Hospital and Tumor Institute, Houston, Texas/ (b) Memorial Sloan-Kettering Cancer Center, New York, USA/ (c) Six main institutions: Brown University, Providence, Rhode Island; St Paul’s Hospital, Vancouver, British Columbia, Canada; UCLA; Houston, Texas; University of Maryland Hospital, Baltimore, Maryland; Memorial Hospital, New York/ (d) Royal Marsden Hospital, London, England/ (e) Hospital A.C. Camargo, Sao Paulo, Brazil/ (f) University of California, San Francisco, USA/ (g) Tata Memorial Hospital, Bombay, India/ (h) Vrije Universiteit University Medical Center, Amsterdam, The Netherlands/ (i) Tom Baker Cancer Centre, Calgary, and Cross Cancer Institute, Edmonton, Alberta, Canada/ (l) Ruhr University, Bochum and Technische University of Munich, Munich, Germany/ (m) St. Luc University Hospital and Cancer Center, Université Catholique de Louvain, Brussels, Belgium/ (n) Negative margin was defined as at least a 1 cm margin of histological clearance.

LRFS: local relapse-free survival; DMFS: distant metastasis-free survival; DSS disease-specific survival; DFS: disease-free survival; OS: overall survival; LRI: incidence of local relapse; DMI: incidence of distant metastasis; CSM: cause-specific mortality. N.S.: not stated.
26%, respectively. These results parallel those reported by major published series [18–32] (Table 3).

Indeed they look quite good, in spite of the difficult location this group of tumors arise from. But if we exclude the relevant proportion of DFSP (35%), which minimally contributed to the occurrence of any events, the outcome becomes much poorer, reflecting the added negative value the “head and neck” site has on prognosis when compared with that of extremity STS [33]. Indeed they look quite good, in spite of the difficult location this group of tumors arise from. But if we exclude the relevant proportion of DFSP (35%), which minimally contributed to the occurrence of any events, the outcome becomes much poorer, reflecting the added negative value the “head and neck” site has on prognosis when compared with that of extremity STS [33]. In fact, 10-year incidence estimates of local relapse, DM and DSM were 27%, 16% and 39%, respectively, when excluding DFSP patients.

Overall, most of the events occurred within the first 3 years after surgery (97% for local relapse, 95% for DM and 83% for cause-specific death). Beyond the third year after surgery, the survival estimates did not substantially change.

As expected, independent prognostic factors for DSM were tumor grade and tumor size; they were also significant for DM and LR, respectively [34].

Of note, at multivariable analysis surgical margins demonstrated only a trend towards an impact on DSM, with borderline significance (P = 0.070). Moreover, no significant association between surgical margins and LR was found. This was possibly affected by the relevant number of DFSP in the positive margins subgroup (17 of 47), 15 of which did not relapse and therefore did neither affect the risk of death.

The prognostic role of the quality of surgery has long been studied in STS. Indeed in extremity STS, an association between surgical margins and survival was consistently shown in several series [3, 35, 36]. In particular, we recently identified a proportion of patients undergoing resection with positive margins, who eventually died of local disease (20%) without developing distant metastases [33]. This high-risk subgroup had a proximally located tumor, which could not be controlled by an amputation. Recent data from our group demonstrated that an improvement in local control may significantly decrease DSM in extremity STS [8]. We hypothesized that this improvement was in part related to the reduction of LR at critical sites, which eventually become incurable. In retroperitoneal STS, local control is even much more important than in extremities, because this site is critical by definition. We recently documented how its improvement can translate into a higher chance of cure [9].

Similarly, the impact of surgical margins on the outcome in HNSTS looks mainly mediated by its impact on LR. In fact when local control is not achieved, tumors may become lethal through local invasion of contiguous vital structures, rather than giving rise to distant spread.

Furthermore, in present series disease-related deaths were almost equally determined by local disease progression and metastatic spread. Mean post-event survival was equally poor for both local and distant recurrence. In other words, in this anatomical region, the impact of LR was as clinically critical as that of metastatic spread.

When looking more deeply to the different HN locations, limited data are as of yet available and no definite conclusion can be drawn on this argument [18, 24, 26, 28].

Our findings support the evidence that sarcomas located both in paranasal sinus and in the supraclavicular region have worst outcome in comparison to other HNSTS. One possible explanation for this finding can be that in these two sites, a higher incidence of high-grade, large and deep tumors were seen (Table 1). Nonetheless, anatomical constraints at these sites (brachial plexus and skull base) may also play an important role.

As far as histology is concerned, leiomysarcoma and MPNST showed the worst outcome. On the contrary, DFSP demonstrated excellent outcome despite the unfavorable anatomical site; overall local control was superimposable to that reported by major published head and neck series [37,38] and comparable with our previous institutional series that included DFSP at any site [39].

Unfortunately, present data are insufficient to evaluate the role of complementary treatments in HNSTS. Nevertheless, a significant proportion of patients with sarcoma other than DFSP received CT and/or RT in the perioperative setting (26 patients out of 108 cases, 24%).

Finally, one more relevant aspect of the series is the number of major reconstructive procedures required (21% of cases), with acceptable morbidity. An extensive use of advanced reconstructive surgery should be considered as an indispensable armamentarium in HNSTS surgery, in order to improve the chance of obtaining adequate surgical margins. Further technical advances are needed, as routine nerve grafting in supraclavicular HNSTS [40].

In conclusion, achievement of wide margins is crucial for cure in HNSTS, since local control could be strictly related to survival. To this end, referral of HNSTS patients to high-volume centers would offer the best availability both of complex demolitive and reconstructive surgical expertise. The specific role of multidisciplinary approach, in particular in the neoadjuvant setting, is worthy of further investigation.

disclosure
The authors have declared no conflicts of interest.

references


