SARCOMAS OF HEAD AND NECK: 31 cases and review of the Literature

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Abstract

Background: Soft tissue sarcomas are a highly heterogenous group of tumors that are classified by differentiation. Their invasive or destructive growth pattern, recurrence and distant metastatic capacity are variable according to their subtype and also their localization. Aim: The aim of this study is to evaluate clinical characteristics, histopathologic subgroups, therapeutic modalities and clinical course of head and neck sarcomas. Study Design: Retrospective study and review of literature Materials and Methods: Patients with head and neck sarcomas who underwent surgery at XXXX University, Faculty of Medicine, Department of Otorhinolaryngology between 2007 to 2016 were retrospectively evaluated. Demographic features of the patients, histopathologic subtypes, immunohistochemical markers, locations of the tumor, type of surgery, margins of the specimens, type of reconstructive surgery and type of adjuvant treatment were reviewed. Results: The histopathologic subtypes were rhabdomyosarcoma, primitive neuroectodermal tumor, osteosarcoma, chondrosarcoma, leiomyosarcoma, Ewing's sarcoma and synovyal sarcoma. The most frequent site was nasal cavity and was detected in 7 cases. Local recurrence developed in three patients. Conclusion: Sarcomas of the head and neck region are relatively rare tumors. Radical resection of the tumors is essential for these cases.

Introduction

Soft tissue sarcomas are a highly heterogenous group of tumors that are classified by differentiation. Their invasive or destructive growth pattern, recurrence and distant metastatic capacity are variable according to their subtype and also their localization. Soft tissue sarcomas are rare with annual incidence 5 per 100000. These tumors are rarely seen in the head and neck region and this region is involved in 4-10 % of soft tissue sarcomas. Only 1-2 % of head and neck tumors are soft tissue sarcomas (1-4). These tumors occur more commonly in males and age and gender properties are variable among the histologic subtypes. Enviromental factors, oncogenic viruses, immunologic factors and genetic predisposition are important in their pathogenesis. The management of these patients requires multidisciplinary approach because of complex anatomy of the head and neck region. Grading and staging systems are important for appropriate evaluation and treatment modalities. Grading system is based on histologic grade, mitotic count, tumor necrosis and tumor differentiation. Staging system requires a multidiciplinary approach again due to the insufficiency of American Joint Committee on Cancer (AJCC) and Musculoskeletal Tumor Society Staging Systems have disadvantages to evaluate these tumors. Clear histopathologic evaluation is essential for the best management. As in many soft tissue sarcomas local control is very important and best treatment is surgery with wide surgical margins for the head and neck sarcomas.

The aim of this study is to evaluate clinical characteristics, histopathologic subgroups, therapeutic modalities

and clinical course of head and neck sarcomas in a single center.

Materials and Methods

Patients with head and neck sarcomas who were operated at XXX University, Faculty of Medicine, Department of Otorhinolaryngology between 2007 to 2016 were retrospectively evaluated in this study. Local ethics committee approval was obtained for this study. The multidisciplinary head and neck team routinely followed all patients; initially 1 month post-operatively and then 3-monthly intervals with computed tomography/Magnetic resonance imaging (CT/MRI), for 5 years or until death. Medical files of patients were evaluated. Demographic features of the patients, histopathologic subtypes, immunohistochemical markers, locations of the tumor, type of surgery, margins of the specimens, type of reconstructive surgery, type of adjuvant treatment were reviewed, Additionally surgical complications, local/regional and distant recurrence patterns and overall survival times were noted.

Results

There were 31 patients; 16 (51,6%) of them were female and 15 (48,4 %) were male. The mean age was 36,2 (range 3-88) years. The histopathologic subtypes were rhabdomyosarcoma, primitive neuroectodermal tumor, osteosarcoma, chondrosarcoma, leiomyosarcoma, Ewing's sarcoma and synovyal sarcoma. The distribution of these sarcoma types has been shown in Figure 1. The mean follow-up was 26,8 monhts (Min:12-Max:96 Months). Tissue samples were firstly examined with hemotoxlylen-eosin. After this evaluation subtyping was performed with imunophenotypic markers including desmin, vimentin, myogenin and TLE-1. Desmin and myogenin were seen possitive an immunohystochemical evaluation of A sample of embrional rhabdomysarcoma which is formed from small-oval and round cells at Figure 2. Chondrosarcoma was seen on the chondromixoid stroma with seperated septa and formed with pleomorphic cells at Figure 3 and TLE1 was seen possitive on an sample of the snovial sarcoma Figure 4.

The most frequent site was nasal cavity and was detected in 7 cases. Other affected sites were maxilla (5 cases), neck (4 cases), parapharyngeal region (1 cases), infratemporal fossa (2 cases), mandible (3 cases), parotid region (2 cases), sphenoid sinus(1 case), malar region (1 case), retrobulber region (2 cases), and larynx (3 cases) (Figure 4). Definitive surgery was performed in 21 cases and 10 patients were inoperable. Total laryngectomy were performed in two patients, total maxillectomy in 6 cases, mandibulectomy and free flap reconstruction in three cases, wide surgical resection in 10 cases. Functional neck dissection was performed in eight cases. Surgical margins were found to be positive in three of the 21 patients who underwent surgical procedures and in 18 cases surgical margins were negative.

Mean follow up was 26,8 months (Min: 12-Max:96 months). Adjuvant chemotherapy was given to 10 cases and radiation treatment to 15 cases. Chemotherapy was consisted of cisplatine, 5-FU, charboplatine, vincristine for adults and vincristine, ifosfamide, Adriamycin, etopocide, ciclofosfamide, charboplatine far pediatric patients. Drugs were chosen according to the patients status.

Local recurrence developed in three patients. One of them was reoperated and chemotherapy was given to the others. Seven of the patients died due to distant metastases.

Discussion

Head and neck region has a unique anatomy that causes variable tumor types originated from muscle, adipous tissue, nerve sheath, vessels and other tissues. Sarcomas are highly heterogenous tumors with different outcome. There are 50 histologic subtypes of sarcoma according to the WHO classification (5). The sarcomas in adults occur 5-15% in the head and neck region (6). The most common sarcomas detected in the head and neck region are osteosarcoma, rhabdomyosarcoma, undifferentiated pleomorphic sarcoma, fibrosarcoma and angiosarcoma(7). The most common sarcoma subtype in head and neck region is rhabdomyosarcoma and this followed by osteosarcoma (7). In our study the most common sarcoma was rahabdomyosarcoma (29 %) and the others were malign mesanchimal tumor, pleomorphic sarcoma, leiomyosarcoma, primitive neuroectodermal tumor, rhabdomyosarcoma, chondrosarcoma, snovial sarcoma, osteosarcoma.

Tumor site is an important factor in decision of surgery. This factor affects the surgical type, clear margins and aesthetic outcomes.

The most commonly seen sarcomas were compatible with the literature. Wide surgical resection with tumor free margins is the best approach for these tumors. Local recurrence rates are high due to rapid growth pattern and high potential for distant metastasis. Multidisciplinary approach is essential for these cases. This approach improve quality of life and cosmetic results and also increases functionality. Primary goal of the treatment of head neck sarcomas is local control. Local control rates are very high in head and neck sarcomas(8). Local control rates were found and has been reported in 47 % to 78 % of the cases(9-10). However overall survival rates are poorer at head and neck region than other locations. Five years overall survival was found between 31 % to 80% in some studies(5,9,10) .

In conclusion, sarcomas of the head and neck region are relatively rare tumors. Local recurrence rates are high. Radical resection of the tumors is essential for these cases. However due to the very high risk of local recurrence and distant metastasis the management of these patients need multidisciplinary approach. These cases must be discussed in multidisciplinary tumor boards and treatment must be individualized according to tumor subtype, localization and patient specific co-morbidities.

References

1.KrausDH.Sarcomasoftheheadandneck.CurrOncolRep 2002;4(1):68–75.

2. FletcherCD.Distinctivesofttissuetumorsoftheheadandneck. ModPathol 2002; 15(3):324–30.

3.WaneboHJ,KonessRJ,MacFarlaneJK,EilberFR,ByersRM,EliasEG,etal.Headandnecksarcoma:reportoftheheadandnecksarcon
of head and neck surgeons committee on research. HeadNeck 1992; 14(1):1–7.

4.ColvilleRJ,CharltonF,KellyCG,NicollJJ,McLeanNR.Multidisciplinary management of head and neck sarcomas. Head Neck 2005;27(9):814–24.

5. Greager JA, Patel MK, Briele HA, Walker MJ, Das Gupta TK. Soft tissue sarcomasof the adult head and neck. Cancer 1985;56(4):820–4.

6. Wanebo HJ, Koness RJ, MacFarlane JK, Elber FR, Byers RM, Elias G, Spiro RH Head and neck sarcoma: report of the head and neck sarcoma registry. Head Neck 1992; 14:1–7.

7. Kraus DH, Dubner S, Harrison LB et al Prognostic factors for recurrence and survival in head and neck soft tissue sarcomas. Cancer 1994;74:697–702

8. Mucke T, Mitchell DA, Tannapfel A, Hölzle F, Kesting MR, Wolff K-D, et al. Out-come in adult patients with head and neck sarcomas—a 10-year analysis. J SurgOncol 2010;102(2):170–4.

9. Chen SA, Morris CG, Amdur RJ, Werning JW, Villaret DB, Mendenhall WM. Adulthead and neck soft tissue sarcomas. Am J Clin Oncol 2005;28(3):259–63.

10. De Bree R, van der Valk P, Kuik DJ, van Diest PJ, Doornaert P, Buter J, et al.Prognostic factors in adult soft tissue sarcomas of the head and neck: a single-centre experience. Oral Oncol 2006;42(7):703–9.

Figure legends

Figure 1: Hystopathologic distrubution of the tumor subtype

Figure 2: Desmin and myogenin were seen possitive an immunohystochemical evaluation of A sample of Embrional Rhabdomysarcoma which is formed from small-oval and round cells.(Desmin:upper, Myogenin:lower)

Figure 3: Chondrosarcoma was seen on the chondromixoid stroma with seperated septa and formed with pleomorphic cells.

Figure 4: TLE1 was seen possitive on an sample of the snovial sarcoma