
Soft Tissue Sarcoma of the Head & Neck: Case Report in Al-Ramadi General Hospital

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Abstract:

Sarcoma in the head & neck area are rare. Management of soft tissue sarcoma in the head & neck is primarily surgical. However the critical anatomy of the head & neck limits the capacity to obtain wide surgical margins. This case report involves a patient who is a 54 years old woman who was referred to the maxillofacial department because of rapidly enlarging mass in the right side involving submandibular space, right cheek and temporal fossa and progressive pain complaint with hearing loss and trismus. Physical examination revealed large firm mass at the right side of the face occupying the submandibular space, infratemporal & temporal fossae, there was trismus, and no other abnormalities were found especially no mucosal lesion or swelling intraorally & no lymph node enlargement in the neck. MRI study was done to the head & neck to study the mass. CT-scan of the chest & abdomen did not show site of distant metastasis. Transoral biopsy through an incision in to the oral vestibule under local anesthesia showed on histopathological examination a malignant peripheral nerve sheath tumor. A surgical resection consisting of parotidectomy, hemimandibulectomy, infratemporal fossa dissection & temporal fossa dissection has been done. Histopathological examination of the surgical specimen revealed positive surgical margins. The patient refused to receive postoperative radiotherapy. Unfortunately 7months postoperatively she developed intracranial metastasis.

Key word: Soft tissue sarcoma

Introduction

Head and neck soft tissue sarcomas are part of a group of malignant soft tissue tumours which can occur in any part of the body containing tissue of mesodermal origin. Soft tissue sarcomas are malignant tumors that may arise in any of the mesodermal tissues of the extremities (50%), trunk and retroperitoneum (40%), or head and neck (10%). Rarely, these tumors arise in the gastrointestinal tract or gastrointestinal stroma, and a small percentage of these are called gastrointestinal stromal tumors (GISTs). Malignant GISTs can occur from the esophagus to the rectum but occur most commonly in the stomach and small intestine. Soft tissue sarcomas occur with greater frequency in patients with [1].

- Von Recklinghausen's disease (neurofibromatosis).
- Gardner's syndrome.
- Werner's syndrome.
- Tuberous sclerosis.
- Basal cell nevus syndrome.
- Li-Fraumeni syndrome (*p53* mutations).

Surgical resection is essential for control of primary sarcoma whether of soft tissue or bone. Although combinations of chemotherapy and radiation may markedly reduce the tumor if given preoperatively, it is essential to remove the full volume as it existed and obtain clear margins. If surgical resection is done prior to planned chemo radiation, clear margins must be obtained. Working closely with the pathologist is important to ensure that gross and frozen section margins are clear [2]. Radiation given preoperatively or post operatively

has been shown to reduce local recurrence. In an older series by Lindberg from M D Anderson, over 300 patients were treated with conservative surgery with removal of gross tumor, but with limited removal of normal tissue (essentially a shell out) and received 60-70Gy of radiotherapy [3].

A standard chemotherapy approach to patients with disseminated sarcomas often involves one of two regimens: (1) doxorubicin (Adriamycin) with or without dacarbazine (DTIC) which has been a standard for many years; or (2) these same two drugs plus ifosfamide (either as the MAID regimen or as A1). In an Intergroup trial the response rate was clearly better with the three-drug MAID regimen than with the two-drug combination of doxorubicin plus dacarbazine (32% vs. 17%) [4,5,6].

The 5 year disease-free survival was 61% for all primary sites and was 63% for head and neck sarcomas. Radiation including brachytherapy can increase local control [7]. The prognosis for patients with adult soft tissue sarcomas depends on several factors, including the patient's age and the size, histological grade, and stage of the tumor [8].

With distant metastases (stage IV), surgery with curative intent is possible for patients selected for optimal underlying biologic behavior (i.e., patients with a limited number of metastases, with a long disease-free interval, and with slow clinical growth) with pulmonary metastases who have undergone or are undergoing complete resection of the primary tumor [9].

Clinical Evaluation:

- Complete history and physical examination includes inspection of surface skin of head and neck with particular attention to skin of scalp and to bone and

soft tissue component of underlying skull and neck^[10,11].

- Complete examination of oropharynx and larynx with attention to possible abnormalities of gum and alveolar ridge and associated soft tissues of mandible/maxilla; inspection via telescope of nasal cavity and nasopharynx^[12].
- Careful evaluation of eye movements, facial skin sensitivity and hearing is important in patients with central facial tumors and facial symmetry and function in patients with lateral skull or retro pharyngeal, preauricular or temporal bone tumors^[13].
- Biopsy of obvious tumor by FNA will indicate general tumor class^[14].
- A confirmatory histological biopsy is necessary. The biopsy report should contain basic information on the histogenetic origin (bone vs. soft tissue) tumor type and grade (high vs. low grade). The ideal report should contain the 6 parameters most often considered including differentiation, cellularity, amount of stroma, vascularity, amount of tumor necrosis and number of mitoses^[15].
- The pathologist should be given full patient information and be informed about x-ray findings especially for bone tumors. Availability of the pathologist in the operation room for special tissue handling may expedite the diagnosis^[16,17].

Presentation of Case

A 54 years old woman was referred to the maxillofacial department because of rapidly enlarging mass in the right side involving submandibular space, right cheek and temporal fossa and progressively increasing pain with hearing loss and trismus.

The patient had been in excellent health until 4 months earlier (at May 2006) when open biopsy of right submandibular lymph node taken and referred for histopathological examination!!!

Microscopical examination of the biopsy specimen revealed lymphatic hyperplasia, CT scan study

before that biopsy (Fig. 1-a) revealed that there is a soft tissue mass irregular in outline, measuring about (3 * 3cm) seen at right submandibular region.

There is bony erosion and sun shine periosteal bone reaction seen at the adjacent right mandibular body due to pressure effect of the mass.

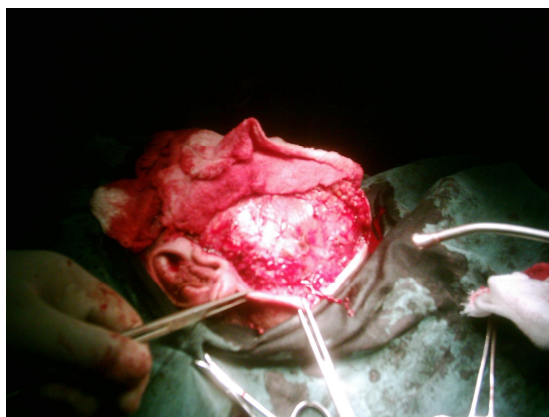
After I.V. contrast media administration (Fig.1-b) the mass shows significant enhancement which is highly suggestive of malignancy.

When the patient reached our department at September 2006 physical examination revealed large firm mass at the right side of the face occupying the submandibular space, infratemporal fossa & temporal fossa, there was trismus, no other abnormalities were found especially no mucosal lesion nor swelling intraorally & there was no lymph node enlargement palpated in the neck.

MRI study (Fig.2-a) showing progression in the mass size about (4*5cm) the mass is isointense & intensity seen engulfing the right mandibular region with effacement of the medial fat plane, the mass causing backward displacement of the right parotid gland. There is central hypo intensity due to central necrosis & calcification. In (Fig.2-b) T2 weighted MRI the mass shows intermediate signal intensity with central hypo intensity (due to calcification) & hypo intensity with central necrosis also there is backward displacement of carotid sheath & medial displacement of fat plane. In the post contrast study –coronal section (Fig.3-a) and (Fig.3-b) the mass shows significant enhancement of the gadolinium contrast media, the mass shows no ipsilateral intracranial extension, no intraorbital nor intraantral extension transoral biopsy through an incision in to the oral vestibule under local anesthesia showed on histopathological examination a malignant peripheral nerve sheath tumor. CT-scan of the chest & abdomen did not show site of distant metastasis. The patient underwent a resection consisting of parotidectomy, hemimandibulectomy, infratemporal fossa dissection & temporal fossa dissection. Histopathological examination of the surgical specimen (Fig.5) revealed positive surgical margins. The patient refused to receive postoperative radiotherapy. Unfortunately 7 months postoperatively she developed intracranial metastasis as seen in CT-scan study. (Fig. 4).



Tracheostomy preoperatively



flap raising intraoperatively



Immediately post operative picture



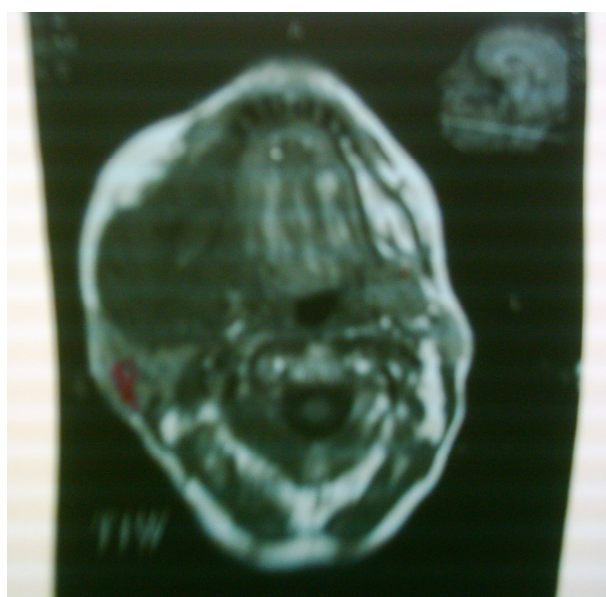
4 months postoperatively



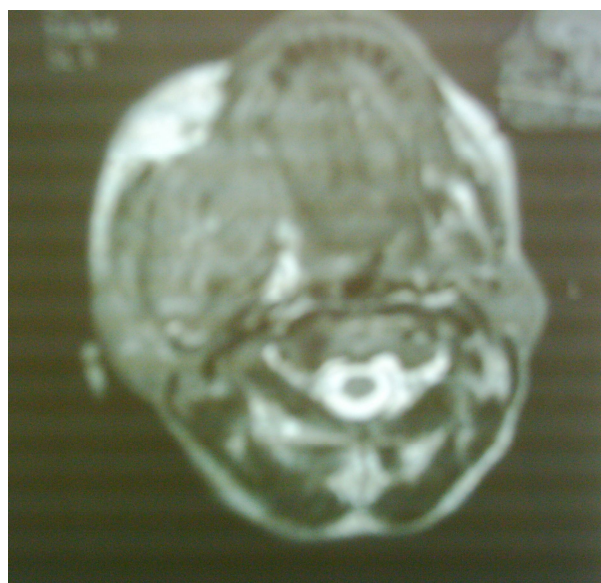


**Irregular soft tissue mass
Fig.1-a**

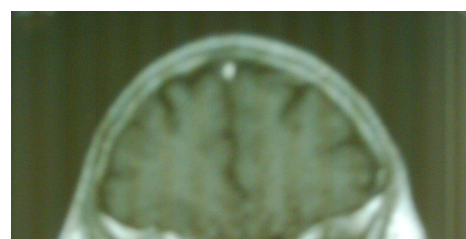
**Enhancement by I.V. contrast
Fig.1-b**

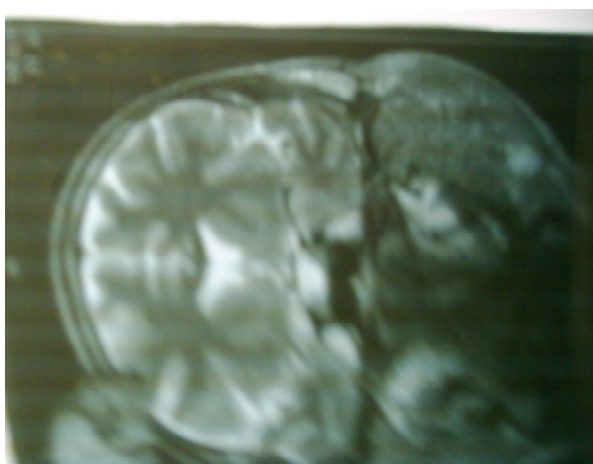


**T1 weighted MRI
Fig.2-a**



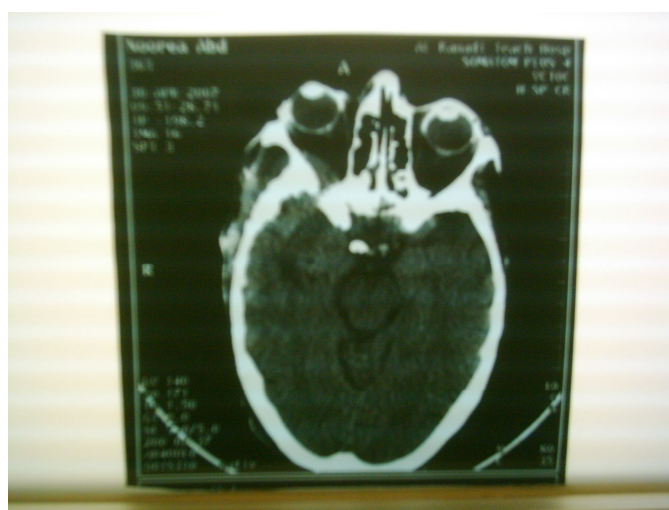
**T2 weighted MRI
Fig.2-b**





Post contrast study –coronal section MRI
Fig.3-a

Fig.3-b



Postoperative CT showing intracranial metastasis
Fig-4



Fig-5
The mass

Discussion

Improper initial management in the form of excisional biopsy of submandibular lymph node taken while ignoring the primary tumor which resulted in delaying the exact diagnosis. Postoperative radiotherapy improves the local control rate, our patient refused it & this increased the risk of recurrence. With delay in the early diagnosis & radical treatment resulted in distant metastasis (brain) as in fig.5 which shows CT-scan study of the brain taken 7 months later for the investigation of unexplained neurological symptoms (headache & fit) starting months post operatively.

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