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Clinical, therapeutic and prognostic features of osteosarcoma of the jaws – Experience of 36 cases

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ABSTRACT

Introduction: Osteosarcoma of the jaws (OSJ) differs from osteosarcoma of other skeletal regions due to later development, a high mortality associated with the local disease, fewer incidences of metastases and its extreme rarity. In regard to clinical and pathological parameters as well as therapeutic approaches and prognosis, OSJ has not been specifically examined to date. In order to achieve a better understanding of this special malignancy, an evaluation of incidence, treatment and prognosis of patients with OSJ in our department over the past 38 years was conducted.
Materials and methods: A retrospective analysis of patients with OSJ between 1972 and 2010 was performed. Information regarding patient characteristics, site of the lesion, main presenting symptoms, latency of initial diagnosis, treatment, histology, local recurrence, development of metastatic disease, duration of follow up and survival was obtained. The data were compared to the literature.
Results: Thirty-six patients (2–81 years, mean: 33.9, standard deviation: 21.3) were diagnosed and treated for OSJ (maxillar:mandibular nearly 1:2). Initial symptoms were local swelling (81%) and pain (47%). The latency period between fist symptoms and clinical presentation was 3.7 months (1–24). A radical resection alone was conducted in 15 patients. In nine patients, resection and radiotherapy was used. Resection with chemotherapy was the treatment of choice in seven patients. Five patients received a triple combination of resection, chemo- and radiation therapy. The osteoblastic subtype of osteosarcoma was most frequent (42%). In 15 cases (42%) local recurrences, in two cases (5%) metastasis were seen. Of these patients, 13 died within the observation period. One other patient (3%) died as a result of progressive pulmonary metastasis. A mean total survival rate of 61% could be seen whereas the highest survival rate (80%) was found in patients who were treated with neoadjuvant chemotherapy, radical resection and adjuvant radiation. Positive prognostic factors were a younger age and tumour-free resection margins.
Discussion: OSJ is a highly lethal tumour entity. According to the data at hand, therapy should possibly include chemotherapy, radical resection and irradiation. Nevertheless, due to the rarity of OSJ, information remains limited and the treatment of choice should be within the focus of clinical multi-centre studies.

1. Introduction

Sarcomas are malignant neoplasms derived from cells of mesenchymal origin. The originating tissue includes bone, cartilage, muscular, fibrous, vascular, fatty and neural tissue (Yamaguchi et al., 2004). Osteosarcoma is attributed to a heterogenous group of primary malignant neoplasms in which mesenchymal cells produce osteoid or immature bone (Amaral et al., 2008; Schajowicz et al., 1995). With an incidence of 0.2–0.3/100,000 per year in Europe, osteosarcoma is the most common malign tumour of the skeletal system (Nathrath et al., 2004). More than half of all osteosarcomas arise in the long bones of the limbs, particularly in the region of knee and pelvis (Dahhn and Coventry, 1967; Garrington et al., 1967). Osteosarcoma of the jaws (OSJ) is uncommon and represents 6–8% of all osteosarcomas (incidence 0.7/1,000,000) (Gadwal et al., 2001; Mardinger et al., 2001). OSJ occurs later than osteosarcoma of the long bones (Batsakis, 1984) with a peak in the second, third and fourth decade (mean age: 35 years (Lee et al.,

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1988)). More men than women are affected (Gadwal et al., 2001; Granowski-LeCornu et al., 2011; Huvos, 1991; Mardinger et al., 2001). Some authors conclude that the most frequent localization is the mandible (Demicco et al., 2010; Prein et al., 1985), whereas others report that the distribution between maxilla and mandible is equal (Saito and Unni, 2005). The aetiological mechanisms are unknown. An influence of chemicals and radiation is reported (Dahlin and Unni, 1977; Franco Gutiérrez et al., 2008) and pre-existing benign bone disorders and trauma were perceived as possible factors in the aetiology of OSJ (Garrington et al., 1967). The development of OSJ in association with dental implant has been reported only once (McGuff et al., 2008). Clinical characteristics are pain and local swelling (Chindia et al., 1998; Nathrath et al., 2004) as well as periapical inflammation and loosening of teeth (Richards and Coleman, 1957). Infiltration of adjacent tissue is common. Dependent on localization, hypothesia and paraesthesia of involved nerves (Nagler et al., 2000), eye symptoms, trismus, nasal obstruction, epistaxis and gingival inflammation can be seen (Batsakis, 1984; Prein et al., 1985; Salvati et al., 1993). Radiographic examination usually shows a lytic, sclerotic or mixed lesion with soft tissue extension in the majority of cases. Further features are widening of the periodontal ligament space and a periosteal reaction (Givol et al., 1998). Craniofacial osteosarcoma does not show explicit pathognomonic signs (Garrington et al., 1967) or differences to osteosarcoma of the rest of the skeleton (Huvos, 1991). For OSJ, a better differentiation than in osteosarcomas of the rest of the skeleton is described (Neville et al., 2002). Chondroplastic osteosarcoma is the most frequent subtype (50% of osteosarcoma of the jaw), followed by osteo- and fibroplastic differentiations (Freyschmidt and Ostertag, 1988). Prognosis is not influenced by the histologic subtype (Granowski-LeCornu et al., 2011; Neville et al., 2002).

OSJ includes various phenotypes (Huvos, 1991). This often complicates the correct diagnosis. The primary difficulty encountered in the diagnosis of jaw lesions is that the clinical features of a number of common dental disorders resemble those of OSJ (Lindqvist et al., 1986). In differential diagnosis, for example simple infections, and diseases such as fibrous dysplasia and osteomyelitis must be taken into consideration (Granowski-LeCornu et al., 2011; Petrikowski et al., 1995). Radiological and, most important, a comprehensive histopathological assessment is necessary (Chindia et al., 1998; Werner et al., 1996).

OSJ have a high tendency towards local relapse (mandible 39–70%; maxilla 15–53%)(Mark et al., 1991; Wilcox et al., 1973). Distant metastasis occur in approximately 18% of the cases (Mardinger et al., 2001; Okinaka and Takahashi, 1997), most frequent in the lung and skeletal system. Locoregional metastasis of lymphatic nodes is rarely seen. Major causes of death by OSJ are complications of uncontrolled local recurrence (Mardinger et al., 2001; Mark et al., 1991). The overall 5-year-survival prognosis is 25–57% (Osteogenic sarcoma of the mandible and maxilla: a Canadian review (1980–2000), 2004; Garrington et al., 1967; Rapidis et al., 2005). An interdisciplinary therapeutic approach of surgery, orthopaedics, pathology, radiology, pre- and post-operative chemotherapy and reconstructive plastic surgery is a prerequisite for the improvement of curing rates (Greenspan, 1993). The benefit of neoadjuvant chemotherapy has not been conclusively evaluated. Hints for a better prognosis when reacting positively on chemotherapy were reported (Jundt and Prein, 2000; Patel et al., 2002), but OSJ seem to react worse to chemotherapy than osteosarcoma of the long bones (Jundt and Prein, 2000). The choice of induction chemotherapy, post-operative chemotherapy or a combination still remains unresolved (Rosenthal et al., 2003).

Due to it's diversity, rarity and obvious differences of OSJ to osteosarcoma of the residual skeletal system, there are also still open questions in regard to clinical and pathological parameters as well as therapeutic approaches and prognosis (Huber et al., 2008). The aim of the present work was to evaluate incidence and prognosis of patients treated for osteosarcoma of the jaws in our department over the past 38 years.

Table 1
Age distribution in relation to diagnosis of osteosarcoma.

Age (years)	Number of osteosarcoma
<10	2
10–19	4
20–29	12
30–39	8
40–49	2
50–59	
60–69	4
70–79	3
>80	1
Total	36

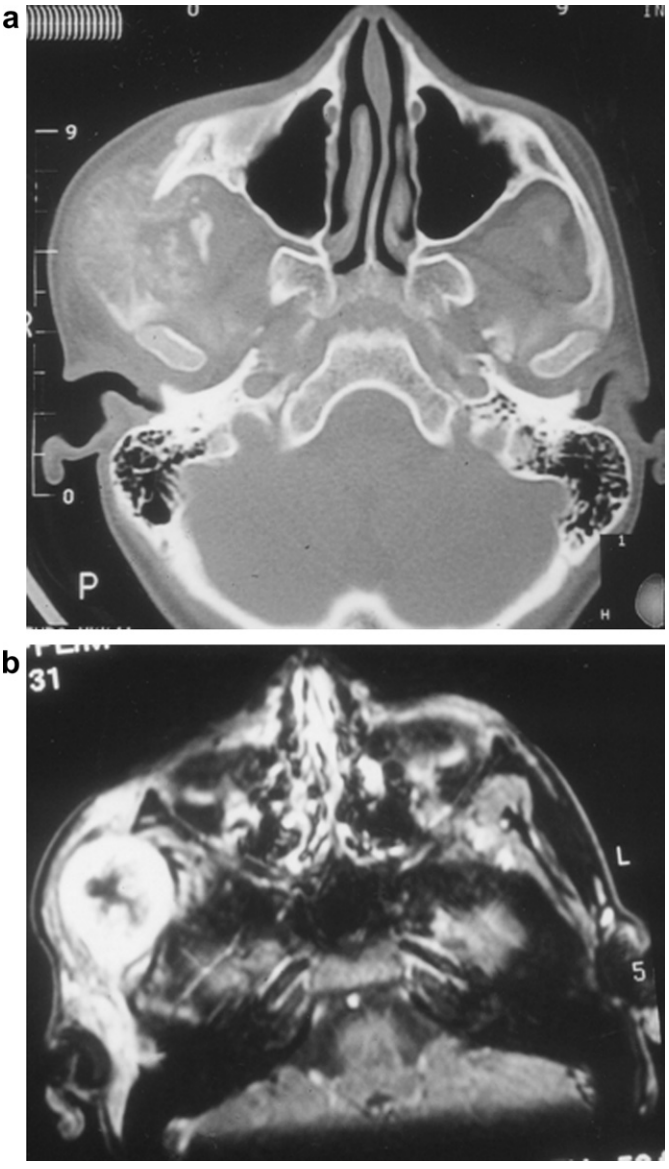

Fig. 1. (a) Computed tomography of an osteosarcoma of the right zygomatic arch. (b) Magnetic resonance tomography (T1) of the same lesion.