Scripta Scientifica Medica, vol. 39 (2007), pp 175-177

MANDIBULAR SARCOMA

Kirov G, Benchev R., Stoyanov St., Kondarev M.

Medical Institute of the Ministry of Interior, Sofia

Reviewed by: Assoc. Prof. K. Ivanov, MD, PhD

ABSTRACT

Sarcomas are comparatively rare tumors – about 1% of all malignancies. They are classified in accordance with the histological results and more than 30 subtypes have been described. 15 to 20% of the sarcomas develop in the maxillofacial region with paranasal cavities and neck being most frequently affected. About 80% of the patient with head and neck sarcoma are adults and 10-20% - children. A clinical case of 31 male with advanced mandibular sarcoma, who was admitted to the Medical Institute of MI after surgery, radio- and chemotherapy is presented. The diagnostic and therapeutic problems of the disease are discussed.

Keywords: sarcoma, mandible, surgery, radiotherapy, chemotherapy.

Sarcomas are comparatively rare tumors, accounting for about 1% of all malignant disease. They are classified in accordance with the histological results, and more of 30 subtypes have been described. 15 to 20% of the sarcomas develop in the maxillofacial area paranasal sinuses and neck being most frequent. About 80% of the patients with head and neck sarcoma are adults, and 10-20% - children.

ETIOPATHOGENESIS

Chondrosarcomas arise in the cartilage or bone tissue and are about 20% from the primary bone tumors. Maxillofacial region is affected in 5 - 10%. The most frequent origin is from the larynx, followed by maxilla, mandible, and scull base. All age groups are affected, but predominates the age 30 – 50 years. About 80% of the chondrosarcomas are primary and develop in healthy tissues, with no clear etiological factor. Secondary lesions are associated with exostosis, Paget's disease; fibrous displasia, Maffucci syndrome or radiation.

CLINICAL PRESENTATION

Chondrosarcomas are slow growing, locally invasive lesions, which comparatively lately give distant metastases. The clinical manifestations are in accordance with the localization. Macroscopically they are hard, lobulated, whitish or bluish submucous lesions. Microscopically can be observed multiple lacunas, containing many pleomorphic nuiclei in a matrix of hyaline cartilage.

Address for correspondence:
Kirov G, Surgical clinic, Medical Institute - Ministry of Interior, 79, Skobelev Blvd, Sofia 1606, BULGARIA

IMAGING

CT can be used for assessment of the degree of bone affection and presence of tumor calcificates, and by means of contrast – for visualization of the near lying vessels. As border areas are not well seen on CT, MRI can be applied for more precise measurements, especially in cases with intracranial spread. Both techniques can be used together, as they give supplemental information. For orientation can be helpful that the malignant chondrosartcomas are usually with diameter greater than 3cm.

TREATMENT

Surgical resection is the method of choice. Regional and distant metastases can be found in less than 10% of the cases at the time of diagnosis. Because of the late metastasis neck dissection is seldom indicated. Local recurrence is the most often cause for failure and can be expected in about 50% of the cases. That's why the radical surgical intervention is extremely important. It is considered that chondrosarcoma are resistant to radiotherapy and in general adjuvant radiotherapy is not recommended except in cases with cranial base involvement, where it can lead to improvement of the lifespan. No clear data exist for improvement after chemotherapy, but it is sometimes applied in poorly differentiated malignancies with distant metastases.

PROGNOSIS

It depends on the degree of differentiation and the localization. Conservative surgery is associated with higher frequency of local recurrence. Poorly differentiated lesions (grade III and IV) are very invasive and have higher rate of

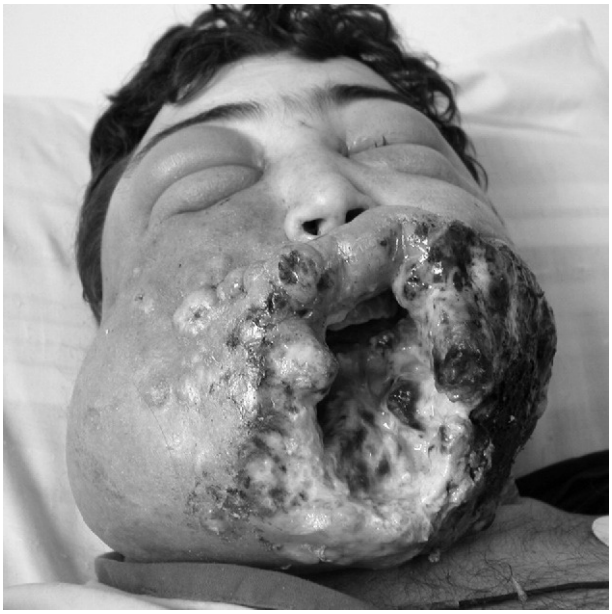
recurrence and metastasis. Distant metastases are 3 times more likely in cases with poor differentiation.

LIFESPAN

5 years lifespan is over 80%, and 10 years – about 70%. Poorly differentiated lesions and cases with distant metastases have lower survival rate. Late appearance of metastases is possible, so longer follow up period (of over 5 years) is recommended.

CLINICAL CASE

We are presenting a clinical case of a 31 years old male with mandibular chondrosarcoma (fig.1,2).



Fif. 1.

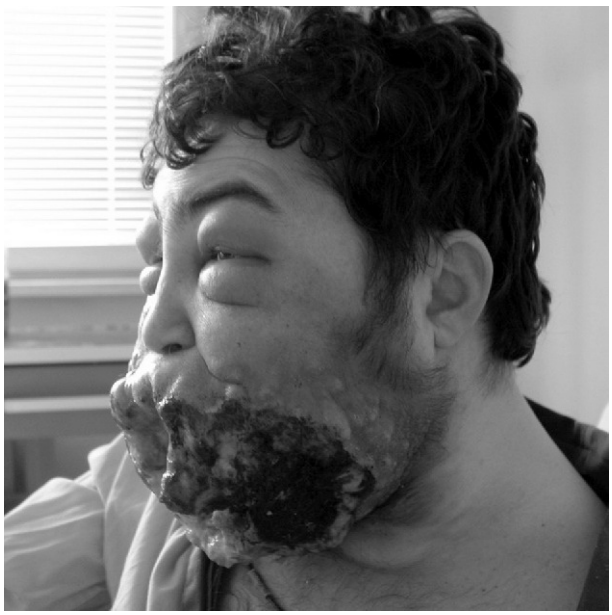


Fig. 2.

The complaints are dating from the beginning of 2003, when first appeared pain and swelling in the right mandibular region. After biopsy has been diagnosed chondrosarcoma and the patient was treated surgically at the HNS of MMA Sofia: in January 2004 was performed radical resection of the mandible and in March 2004 autotransplantation with rib cartilage. In January 2005 a swelling with progressive growth appeared again, which gradually affected the right mandibular, buccal and submandibular regions. In the clinic of oncology of MHAT „St. Marina" consequently were undertaken 3 courses of chemotherapy. In November 2005 the patient was admitted at the Oncology clinic of University hospital Copenhagen where a course of radiotherapy is applied with a result "partial regression of the tumor". From 27.02.2006 till 08.03.2006, because of severe furunculosis in the region of the tumor, the patient was hospitalized at the Department of Dermatological and Venereal Diseases of Medical Institute – Ministry of Interior. He has been dismissed with improvement of the dermatological symptoms after the antibiotic treatment. On 09.04, the patient was admitted at MHAT „St. Ivan Rilski", Razgrad, because of new ulceration of the skin in the region of the tumor with abundant suppuration. On 16.04, he was transferred to MI – MI, ICU.

OBJECTIVELY AT THE ADMISSION

Male in impaired general state. Adequate. Febrility 37.5°C. Severe edema of the left facial scull. Big tumor formation, engaging the lower 2/3 of the face with livid coloration, in advanced ulceration (fig.3). Heart, lungs and viscera – normal. Decubital ulcer in the left gluteal region.

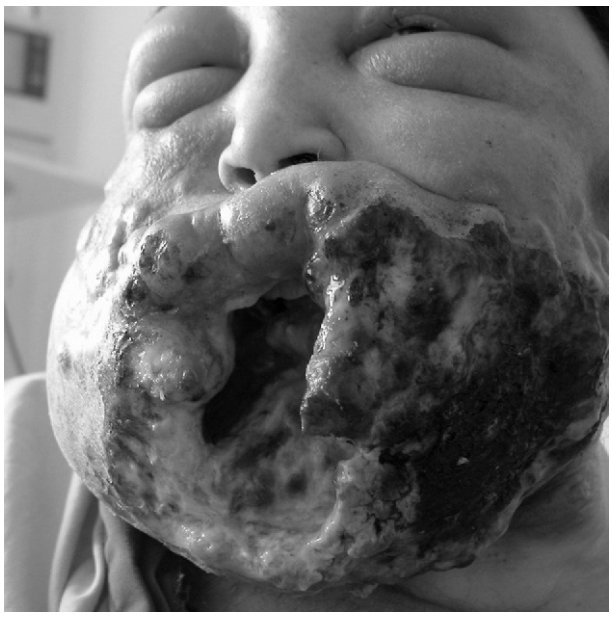


Fig. 3.

On CT, in the right mandibular region is seen big formation with bone density and irregular structure (fig.4,5). The ad-

jacent soft tissues are infiltrated and deformed. Cervical structures are without pathological changes. No enlarged cervical or mediastinal lymph nodes are detected. Chest wall and pleura – unaffected. Localized pneumofibrous changes in the base of the left lung.

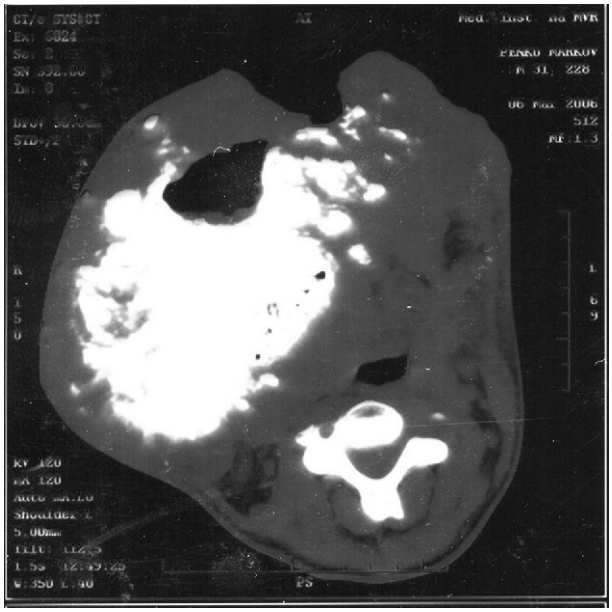
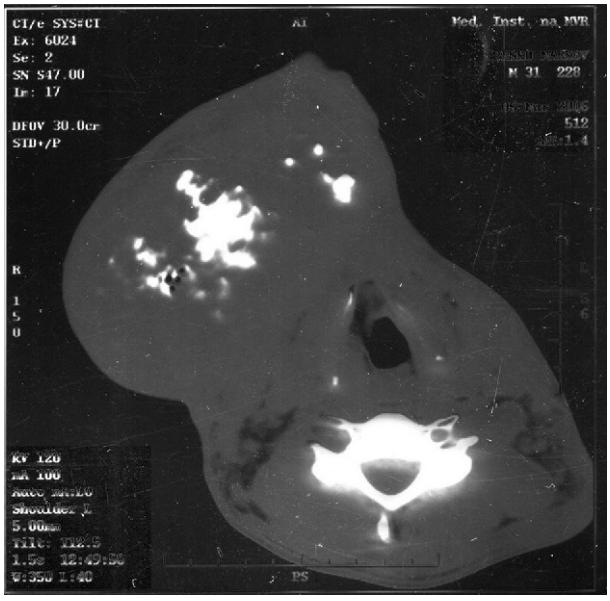


Fig. 4.



After reference to the National consultant of HNS, was taken decision for palliative treatment, as due to the advanced stage of the disease, no radical intervention was possible. On 17.04. after development of acute cardiovascular deficiency, the patient died.

CONCLUSIONS

Head and neck sarcomas are tumors with outlined local invasiveness. For their successful treatment are important the early diagnosis and maximally radical surgical intervention.

The metastases are late, but the percentage of local recurrence is high. In cases when plastic reconstruction is necessary it should be postponed till good tumor control is achieved. So far there are no clear clinical data for the influence of radio- and chemotherapy on the survival rate of the patients.

REFERENCES

1. Colreavy MP, Lacy PD, Hughes J, et al: Head and neck schwannomas - a 10 year review. *J Laryngol Otol* 2000 Feb; **114**(2): 119-24.
2. Daya H, Chan HS, Sirkin W, Forte V: Pediatric rhabdomyosarcoma of the head and neck: is there a place for surgical management? *Arch Otolaryngol Head Neck Surg* 2000 Apr; **126**(4): 468-72.
3. del Rosario ML, Saleh A: Preoperative chemotherapy for congenital hemangiopericytoma and a review of the literature. *J Pediatr Hematol Oncol* 1997 May-Jun; **19**(3): 247-50.
4. Evans HL, Ayala AG, Romsdahl MM: Prognostic factors in chondrosarcoma of bone: a clinicopathologic analysis with emphasis on histologic grading. *Cancer* 1977 Aug; **40**(2): 818-31.
5. Inoue YZ, Frassica FJ, Sim FH, et al: Clinicopathologic features and treatment of postirradiation sarcoma of bone and soft tissue. J *Surg Oncol* 2000 Sep; **75**(1): 42-50.
6. Koch BB, Karnell LH, Hoffman HT, et al: National cancer database report on chondrosarcoma of the head and neck. *Head Neck* 2000 Jul; **22**(4): 408-25.
7. Moley JF, Eberlein TJ: Soft-tissue sarcomas. *Surg Clin North Am* 2000 Apr; **80**(2): 687-708.
8. Oda D, Bavisotto LM, Schmidt RA, et al: Head and neck osteosarcoma at the University of Washington. *Head Neck* 1997 Sep; **19**(6): 513-23.
9. Patel SG, Shaha AR, Shah JP: Soft tissue sarcomas of the head and neck: an update. *Am J Otolaryngol* 2001 Jan-Feb; **22**(1): 2-18.
10. Patel SG, See AC, Williamson PA, et al: Radiation induced sarcoma of the head and neck. *Head Neck* 1999 Jul; **21**(4): 346-54.
11. Sheppard DG, Libshitz HI: Post-radiation sarcomas: a review of the clinical and imaging features in 63 cases. *Clin Radiol* 2001 Jan; **56**(1): 22-9.
12. Smeele LE, Snow GB, van der Waal I: Osteosarcoma of the head and neck: meta-analysis of the nonrandomized studies. *Laryngoscope* 1998 Jun; **108**(6): 946.
13. Wanebo HJ, Koness RJ, MacFarlane JK, et al: Head and neck sarcoma: report of the Head and Neck Sarcoma Registry. Society of Head and Neck Surgeons Committee on Research. *Head Neck* 1992 Jan-Feb; **14**(1): 1-7.