Kulak Burun Bogaz Ihtis Derg
2003;11(5):144-147

CASE REPORT

A case of Ewing's sarcoma in the mandible and the skull base

Mandibula ve kafa tabanında Ewing sarkomu: Olgu sunumu

M. Tayyar KALCIOĞLU, M.D.,[1] Semih ÖNCEL, M.D.,[2] Murat Cem MİMAN, M.D.,[1]
Tamer ERDEM, M.D.,[1] Bülent MIZRAK, M.D.[3]

A thirteen-year-old boy with Ewing's sarcoma was presented. The treatment was considerably delayed because of the initial diagnostic difficulties due to its insidious presentation and the unwillingness of the child's parents to further investigations and treatment. During a three-year delay, the tumor turned out to be a gross painful mass from a painless lesion. Cranial computed tomography and magnetic resonance imaging with contrast showed a destructive lesion extensively involving the right mandible and the skull base, with invasion to the intracranial space. Facial and intratemporal portions of the mass were removed totally, but the intracranial extension could not be totally excised because of cavernous sinus involvement. Histologic and immunohistochemical findings were consistent with the diagnosis of Ewing's sarcoma. The patient received systemic chemotherapy and radiotherapy following surgery. A month after radiation therapy, radiologic investigations showed a lung mass suggestive of metastasis. He died two days after hospitalization, from an intracranial hemorrhage associated with the intracranial mass.

Key Words: Child; combined modality therapy; head and neck neoplasms; prognosis; sarcoma, Ewing's/diagnosis/surgery/radiography/chemotherapy.

On üç yaşında bir erkek çocukta Ewing sarkomuna rastlandı. Tedavi, tümörün başlangıçta az semptom vermesinden kaynaklanan tanı güçlükleri ve hastanın anne-babasının daha ileri araştırma ve tedaviyi kabul etmemeleri nedeniyle önemli ölçüde gecikti. Yaklaşık üç yıllık bir gecikmeden sonra, tümörün ağrısız bir lezyondan ağrılı büyük bir kitleye dönüştüğü görüldü. Bilgisayarlı tomografi ve kontrastlı manyetik rezonans görüntülerinde sağ mandibula ve kafa tabanını yaygın olarak tutan ve kafa içi uzanım gösteren tümör izlendi. Lezyonun yüzdeki ve intratemporal bölümleri tümüyle çıkarılırken, kafa içi uzanımı, kavernöz sinüs tutulumu yüzünden tümüyle çıkarılamadı. Histolojik ve immünkimyasal bulgular Ewing sarkomu tanısını destekler nitelikteydi. Hastaya cerrahi tedaviden sonra sistemik kemoterapi ve radyoterapi uygulandı. Radyoterapiden bir ay sonraki radyolojik incelemelerde akciğerde metastaz düşündüren bir kitle saptandı. Hasta, yatırılmasından iki gün sonra, kafa içi kitleye bağlı gelişen intrakranyal kanama nedeniyle yaşamını yitirdi.

Anahtar Sözcükler: Çocuk; kombine tedavi; baş-boyun neoplazileri; prognoz; sarkom, Ewing/tanı/cerrahi/radyoterapi/kemoterapi.

Ewing's sarcoma was described by James Ewing in 1921.[1] Of the most common primary malignant tumors of bone found in children, it is the second primary malignancy following osteosarcoma. Ewing's sarcoma is an aggressive tumor with poor differentiation, commonly arising in the diaphyses of long bones, ribs, pelvis and vertebrae at the ages of 10 to 15 years.[2,3] Primary localization in the face is very rare and occurs in only 1% to 4% of cases.[3] There are no pathognomonic clinical findings for

◆ *Departments of [1]Otolaryngology and [3]Pathology, Medicine Faculty of İnönü University, Malatya; [2]Department of Otolaryngology, Atatürk State Hospital, İzmir, Turkey.*

◆ *Received: October 10, 2002. Accepted for publication: April 28, 2003.*

◆ *Correspondence: Dr. M. Tayyar Kalcıoğlu. İnönü Üniversitesi Tıp Fakültesi, KBB Hastalıkları Anabilim Dalı, 44300 Malatya, Turkey. Tel: +90 422 - 341 06 60 / 4607 Fax: +90 422 - 341 07 28 e-mail: mtkalcioglu@hotmail.com*

* *Presented at the 1st Joint Meeting of Otorhinolaryngology, Head and Neck Surgery Societies of Turkey and Greece (Sep 22-26, 2001, Antalya, Turkey).*

◆ *İnönü Üniversitesi Tıp Fakültesi, [1]Kulak Burun Boğaz Hastalıkları Anabilim Dalı, [3]Patoloji Anabilim Dalı, Malatya; [2]İzmir Atatürk Eğitim Hastanesi Kulak Burun Boğaz Hastalıkları Kliniği, İzmir.*

◆ *Dergiye geliş tarihi: 10 Ekim 2002. Yayın için kabul tarihi: 28 Nisan 2003.*

◆ *İletişim adresi: Dr. M. Tayyar Kalcıoğlu. İnönü Üniversitesi Tıp Fakültesi, KBB Hastalıkları Anabilim Dalı, 44300 Malatya. Tel: 0422 - 341 06 60 / 4607 Faks: 0422 - 341 07 28 e-posta: mtkalcioglu@hotmail.com*

* *1. Türk-Yunan Otorhinolarengoloji ve Baş-Boyun Cerrahisi Dernekleri Ortak Toplantısı'nda sunulmuştur (22-26 Eylül 2001, Antalya).*

Ewing's sarcoma in the head and neck region. The most common manifestations arise from the mass effect of the tumor. It presents as a slow growing, hard mass associated with mild tenderness or pain. Loosening of the teeth, local paresthesia, or otitis media may occur.

CASE REPORT

A thirteen-year-old boy presented with a painless mass of a year duration in the right parotid region. He did not have any other symptoms. He had undergone fine-needle aspiration biopsy at another center a year ago and the result had been reported as benign. After a month of his presentation to our center, superficial parotidectomy, together with an excisional biopsy was performed.

Pathological examination was reported as a low-grade neuroectodermal tumor, requiring immunohistochemical study for classification. However, his parents accepted neither the immunohistochemical analysis of the specimens nor further treatment. The patient was lost to follow-up for about three years, after which he presented with a complaint of pain and with a gross mass covering 1/3 of the right facial half. Routine laboratory examinations were normal. Cranial computed tomography and magnetic resonance imaging with contrast revealed a destructive lesion extensively involving the right mandible and the skull base, with invasion to the intracranial space (Fig. 1a, b).

An excisional biopsy was planned with the department of neurosurgery. Facial and intratemporal portions of the mass were removed totally, but the intracranial extension could not be totally excised because of cavernous sinus involvement. The defects in the cranium and the skull base were reconstructed by methyl methacrylate. Histologic examination revealed a small cell tumor with a round nucleus. Periodic acid-Schiff staining was highly positive (Fig. 2a). Immunohistochemical staining was found positive for chromogranin A antibody (Fig. 2b), but negative for leukocyte common antigen, cytokeratin, and desmin. These findings were all consistent with a diagnosis of Ewing's sarcoma. Evaluation for metastatic disease was negative. Systemic chemotherapy was started because of local control of disease and to prevent systemic micrometastasis. The treatment protocol consisted of cyclophosphamide, vincristine, Adriamycin, and dactinomycin as proposed by Burgert et al.[4] After the completion of three courses of treatment, radiation therapy with a total dose of 6.000 cGy was

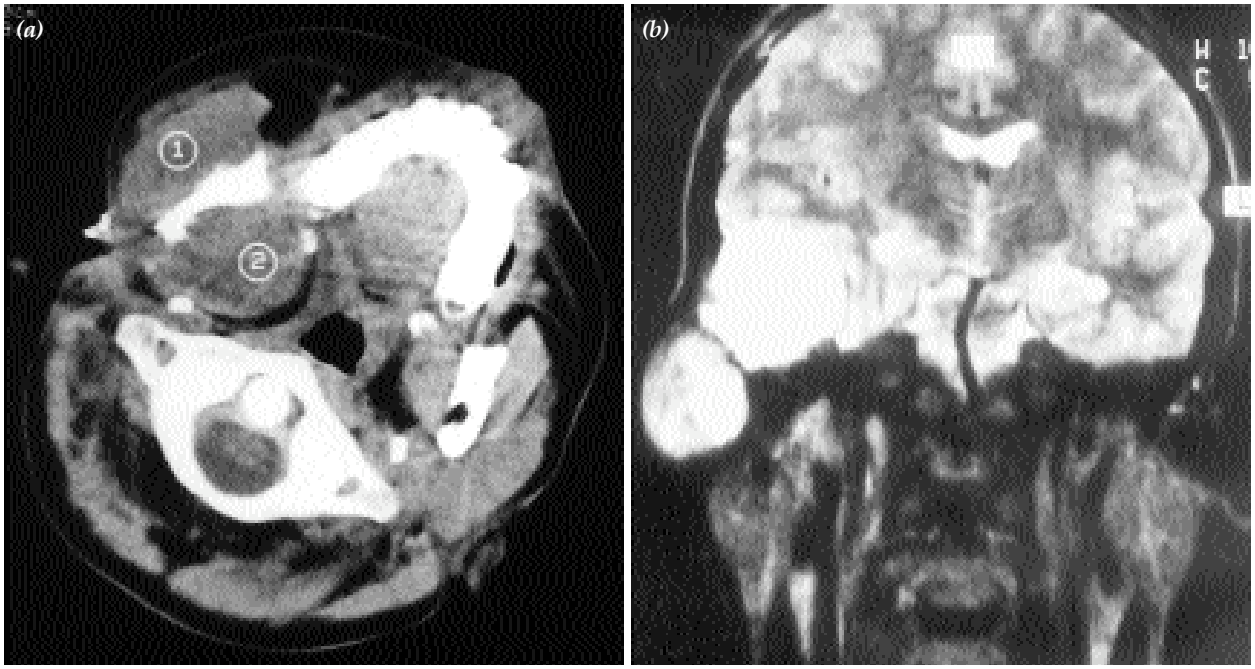

Fig. 1 - ***(a)*** *Axial computed tomography showing a destructive lesion extensively involving the right parotid and the mandible.* ***(b)*** *T_2-weighted coronal magnetic resonance image showing a hyperintense mass involving the skull base, with invasion to the intracranial space.*

administered. However, local disease was detected a month after radiation therapy. Radiologic investigations showed a lung mass suggestive of metastasis. He died two days after hospitalization from an intracranial hemorrhage associated with the intracranial mass.

DISCUSSION

Ewing's sarcoma is rare in the head and neck region; its frequency in bones of the head and neck accounts for about 1% to 4% of all Ewing's sarcoma cases.[3,5,6] Primary bone tumors of this region arise most commonly from the skull, the mandible, and the maxilla.[3] The tumor is usually slow growing and hard, accompanied by tenderness or dull pain. Periosteal reaction, cortical thickening or soft tissue masses can be seen on radiologic examinations; however, other manifestations such as lytic changes and cortical violation are more evident.[3] Therefore, diagnosis can easily be mistaken for an inflammatory disease, causing delays in the institution of correct treatment.[2] On the first presentation of this patient, radiologic findings suggested only the presence of a soft tissue mass without any bone defect. In addition, he had undergone an aspiration biopsy, which failed to show any malignant cells. After

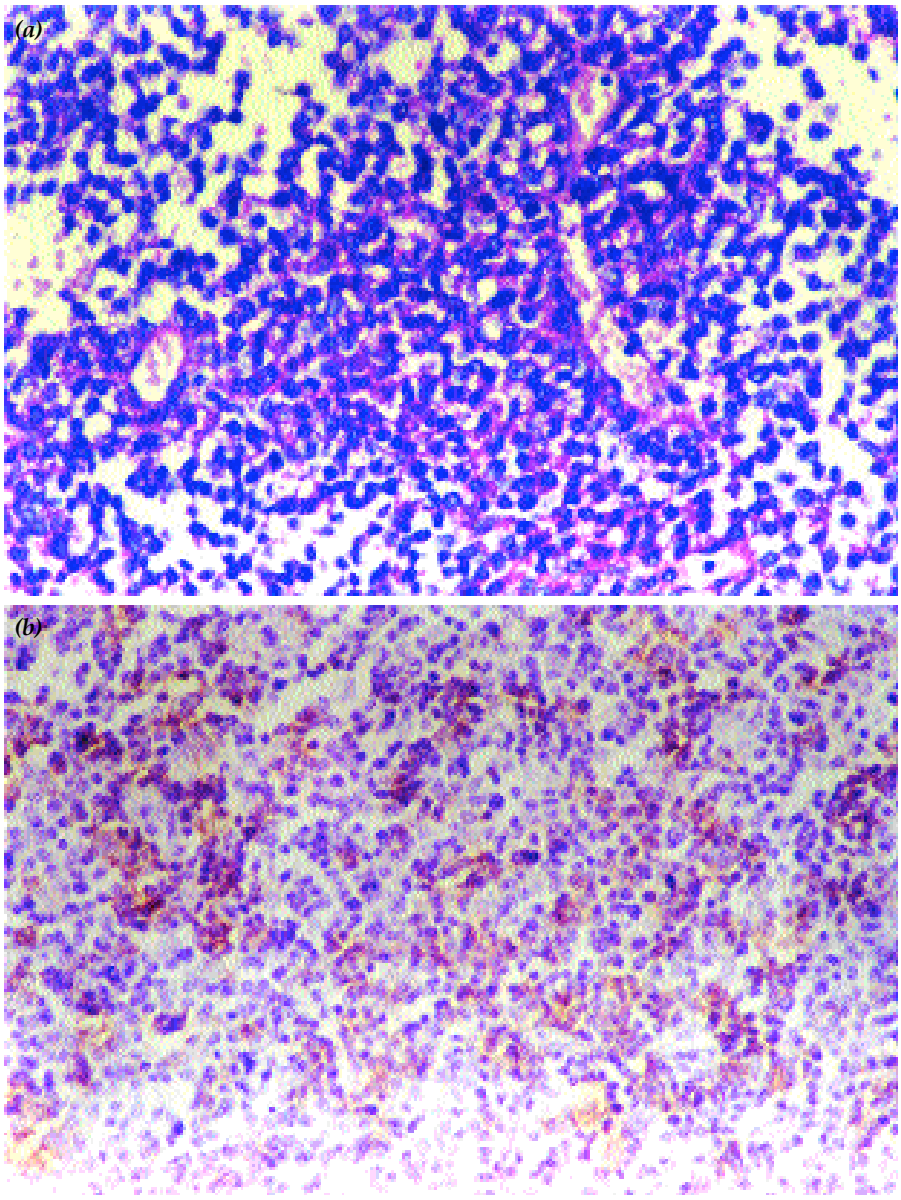

Fig. 2 - ***(a)*** *Diffuse cytoplasmic positive staining of monotonous round cells (PAS x 50).* ***(b)*** *Patchy positive staining of tumor cells for chromogranin A antibody (Streptavidin-biotin technique with diaminobenzidine as a chromogene x 50).*

superficial parotidectomy, reaching a precise diagnosis and implementing an appropriate treatment were hampered by his parents' unwillingness to cooperation.

Although patients with axial localization show very poor prognosis,[6-8] some authors reported improved prognosis in Ewing's sarcomas involving the head and neck.[2,3,5] The five-year survival rate was found as 68% with combination surgery followed by radiation therapy, compared with 54% and 43% with surgical excision and primary radiation therapy, respectively.[1] Daw et al.[5] reported a survival rate of 33% in patients treated with complete or incomplete surgical resection and radiotherapy. The survival was about five years in our case, despite incomplete investigations and insufficient treatment from the first appearance of symptoms.

Surgical treatment, radiation therapy, or chemotherapy can be used separately or in combination for the treatment of Ewing's sarcoma. Daw et al.[5] employed combined therapy in seven patients; of these, two patients underwent complete resection, and five patients underwent incomplete resection. They observed a better prognosis following complete resection.

Radiotherapy is almost always required,[5] although there are some authors who find it unnecessary after complete resection of primary tumors.[9] The efficiency of radiation therapy has been demonstrated in several studies.[8,10] Fiorillo et al.[2] reported improved results after high dose radiotherapy.

Treatment with chemotherapy is essential for the control of metastatic disease and to enhance the possibility of local control.[5] Systemic therapy is necessary because of a very high incidence of occult micrometastases.[8] Therefore, it should include both local and systemic therapies. It may improve the disease-free survival rate.[1] The survival rate was reported as 68% in patients receiving a high-dose systemic therapy with vincristine, Adriamycin, and cyclophosphamide.[4]

In conclusion, it must be kept in mind that Ewing's sarcoma of the head and neck can easily be mistaken for an inflammatory disease. At the beginning, it may appear a soft tissue mass, without any apparent bone defects. A combination of complete surgical therapy, radiotherapy, and chemotherapy may prolong the survival.

REFERENCES

1. Vaccani JP, Forte V, de Jong AL, Taylor G. Ewing's sarcoma of the head and neck in children. Int J Pediatr Otorhinolaryngol 1999;48:209-16.
2. Fiorillo A, Tranfa F, Canale G, Fariello I, D'Amore R, De Chiara C, et al. Primary Ewing's sarcoma of the maxilla, a rare and curable localization: report of two new cases, successfully treated by radiotherapy and systemic chemotherapy. Cancer Lett 1996;103:177-82.
3. Siegal GP, Oliver WR, Reinus WR, Gilula LA, Foulkes MA, Kissane JM, et al. Primary Ewing's sarcoma involving the bones of the head and neck. Cancer 1987; 60:2829-40.
4. Burgert EO Jr, Nesbit ME, Garnsey LA, Gehan EA, Herrmann J, Vietti TJ, et al. Multimodal therapy for the management of nonpelvic, localized Ewing's sarcoma of bone: intergroup study IESS-II. J Clin Oncol 1990;8:1514-24.
5. Daw NC, Mahmoud HH, Meyer WH, Jenkins JJ, Kaste SC, Poquette CA, et al. Bone sarcomas of the head and neck in children: the St Jude Children's Research Hospital Experience. Cancer 2000;88:2172-80.
6. Wood RE, Nortje CJ, Hesseling P, Grotepass F. Ewing's tumor of the jaw. Oral Surg Oral Med Oral Pathol 1990; 69:120-7.
7. Rosen G, Caparros B, Nirenberg A, Marcove RC, Huvos AG, Kosloff C, et al. Ewing's sarcoma: ten-year experience with adjuvant chemotherapy. Cancer 1981; 47:2204-13.
8. Dunst J, Sauer R, Burgers JM, Hawliczek R, Kurten R, Winkelmann W, et al. Radiation therapy as local treatment in Ewing's sarcoma. Results of the Cooperative Ewing's Sarcoma Studies CESS 81 and CESS 86. Cancer 1991;67:2818-25.
9. Hayes FA, Thompson EI, Meyer WH, Kun L, Parham D, Rao B, et al. Therapy for localized Ewing's sarcoma of bone. J Clin Oncol 1989;7:208-13.
10. Bacci G, Picci P, Gitelis S, Borghi A, Campanacci M. The treatment of localized Ewing's sarcoma: the experience at the Istituto Ortopedico Rizzoli in 163 cases treated with and without adjuvant chemotherapy. Cancer 1982;49:1561-70.