CASE REPORT

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Isolated maxillary sinus Ewing's sarcoma*

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SUMMARY

Extraskeletal Ewing's sarcoma is rarely found arising in the head and neck region. Only six cases of extraskeletal Ewing's sarcoma invading the nasal cavity or paranasal sinuses have been reported in the literature. The first described case of that Ewing's sarcoma located in isolated maxillary sinus extending into the orbita without involving the nose is presented. A 16-year-old woman presented with swelling in her right cheek. The nasal examination was normal. A computed tomography (CT) and magnetic resonance imaging (MRI) of the sinuses revealed a large mass in the right maxillary sinus with extension to the orbita. Under general anesthesia biopsies were taken. Based on histopathological analysis, a diagnosis of extraskeletal Ewing's sarcoma was made. The patient did not accept the surgical therapy. The patient was treated with combined chemotherapy and radiotherapy. The patient has done without evidence of recurrence or metastasis for one year.

Key words: Ewing's, sarcoma, extraskeletal Ewing's sarcoma, maxillary sinus tumour, nose neoplasm

INTRODUCTION

Ewing sarcoma, the most frequent small and round cell bone tumour of early childhood, childhood and adolesence and rarely of adulthood, has two types: the classical skeletal type and the extraskeletal type [1, 2]. An even rarer subset of Ewing's sarcoma, known as the extraskeletal Ewing's sarcoma, arises from soft tissue, rather than in relationship to bone. Extraskeletal Ewing's sarcoma shares similar histological, immunohistochemical and molecular findings with Ewing's sarcoma of bone [3, 4]. The head and neck have seldom been involved as primary site of the extraskeletal type, the majority occur in the mandible or the maxilla [3].

Only six cases of exstraskeletal Ewing's sarcoma involving the nasal cavity and / or paranasal sinuses have been reported in the literature [3]. A case of a maxillary sinus Ewing's sarcoma in a 16-year-old patient is discussed as the first described case of this tumour isolated in maxillary sinus without involvement of the nose.

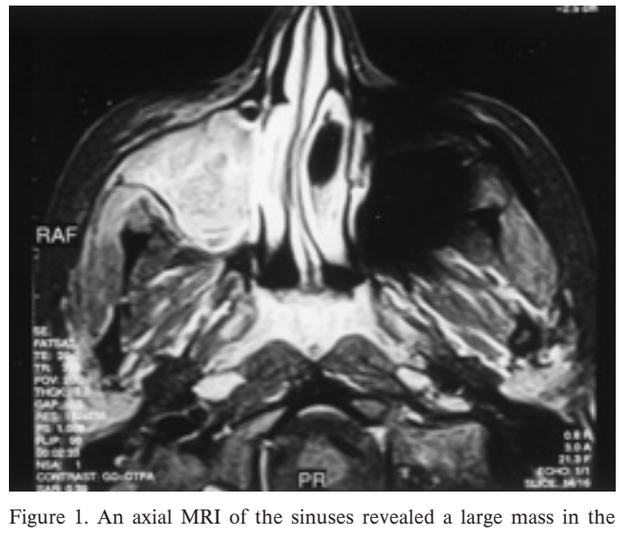

Figure 1. An axial MRI of the sinuses revealed a large mass in the right maxillary sinus.

CASE REPORT

A 16-year-old girl presented with a non-tender enlargement in her right cheek for 4 months. She didn't complain of rhinorrhea, nasal obstruction or epistaxis. On examination a firm swelling was noted in her right cheek. There was no diplopia. Fiberoptic examination revealed a normal nasal cavity. A magnetic resonance imaging and computed tomography scan of the sinuses revealed a large mass in the right maxillary sinus with an extension to the orbita (Figures 1 and 2). There was erosion in the inferior orbital wall but there wasn't a contiguous spread. The remaining sinuses, orbital contents and cranial fossa were uninvolved. The patient was taken to the operating room and underwent a Caldwell-luc operation as a biopsy. The mass was large, lobulated and vascular. Based on histological

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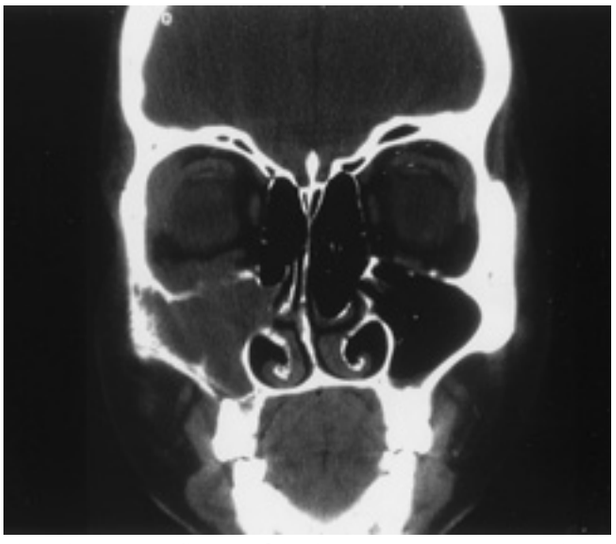

Figure 2. A coronal CT scan of the sinuses revealed a large mass in the right maxillary sinus with an extension to orbita.

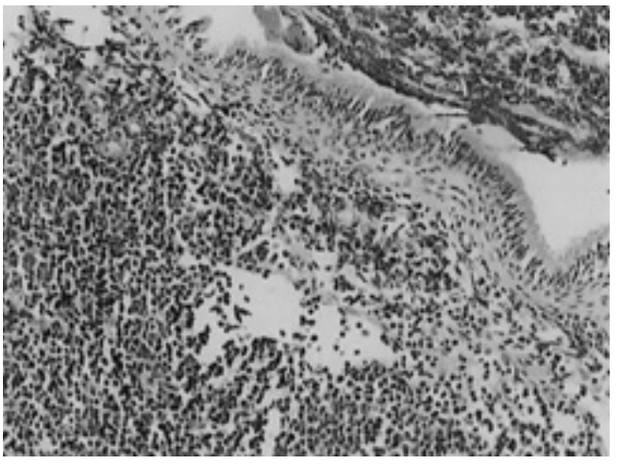

Figure 3. The tumour cell groups are separated by fine bands. The tumour cells have uniform, round or oval nucleus with dispersed chromatin inconspicuous nucleoli and scanty cytoplasm (HEx400).

analysis a diagnosis of Ewing's sarcoma was made. An extensive metastatic work-up postoperatively was negative; this included CT scans of the chest, abdomen, and MRI of the brain and bone scanning. Microscopic analysis revealed a tumour consisting of small-round blue cells with pale cytoplasm and oval nuclei. The tumour consisted of a solid sheet of small, round blue cells (Figure 3). Mitotic figures were present as well as individual cell necrosis. It was locally richly vascularized. A periodic acid-schiff stain demonstrated diastase sensitive glycogen in the cytoplasm of many cells. Multiple immunohistochemical stains of the specimen were performed. From these stains the most diagnostic was the CD99 stain which was strongly positive (Figure 4). The neoplastic cells reacted slightly with NSE, and focally with CD3. No

Table 1. Literature review of the Extraskeletal Ewing's sarcomas of the head and neck region.

Author	Sex	Age year	Localization	Therapy	Histochemical Findings	Immunhisto chemical Analysis	Electron Microscopy Findings
Pontius et al., 1981	M	39	L nasal fossa	Craniofacial resection + RT	PAS + (Glycogen)		Glycogen rosettes
Lane et al., 1990	M	7	R nasal cavity, ethmoid sinus, orbita	Partial intranasal ethmoidectomy	PAS + (Glycogen)	Vimentin +	Glycogen
Howard et al., 1993	M	14	R L ethmoid sinuses, R nasal cavity, Med. orbital wall, cribriform plate	Craniofacial resection + CT + RT	Reticulin +		Glycogen rosettes
	F	28	Nose	Partial rhinectomy + RT + CT	Reticulin +		Glycogen rosettes
Aferzon et al., 2000	F	14	L and ethmoid complex	Ant + Post ethmoidectomy + CT + RT		CD99 +	
Böör et al., 2001	F	20	Nose	Surgery + CT + RT	Glycogen +	CD99 + Vimentin +	
Uslu et al., 2003	F	16	R maxillary sinus + orbita	CT + RT	PAS + (Glycogen)	CD99 +	

F: Female, M: Male, L: Left, R: Right, CT: Chemotherapy, RT: Radiotherapy

immunoreactivity was observed with CD20, CD30, synaptohysin, chromogranin, desmin and LCA. Surgery has been proposed to the patient but she did not accept it. Afterwards the patient was treated with combined chemotherapy / radiotherapy. The adjuvant chemotherapy program, consisted of treatment every 3 weeks with vincristine, doxorubicin, cyclophosphamide, dactinomycin alternating with ifosfamid and etoposide through week 33 (17 cycle's total) [5]. At week 12, the patient received radiation therapy for local control (total dose: 5040 cGy in 28 fractions). The patient has done well without evidence of recurrence or metastasis for one year.

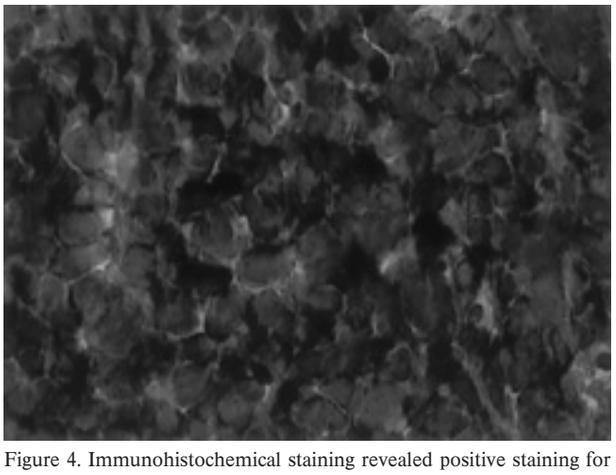

Figure 4. Immunohistochemical staining revealed positive staining for CD99 (x400).

DISCUSSION

Ewing's sarcoma is a malignant neoplasm of uncertain histogenesis which most commonly arises in the skeleton [6]; however extraskeletal Ewing's sarcoma and round cell soft tissue sarcomas fulfilling the diagnostic criteria for Ewing's sarcoma are recognized [7]. The head and neck are unusual sites for primary skeletal Ewing's sarcoma and most described cases in these locations have arisen from the maxilla [8]. There are 6 cases of Ewing's sarcoma involving the nasal cavity and paranasal sinuses [3]. Ewing's sarcoma may originate from bone or, less frequently, from soft tissues. The demarcation of the extraskeletal soft tissue type from the classical skeletal type is practically impossible [9]. In the WHO classification the soft tissue Ewing's sarcoma has been regarded as identical with the osseous analogue [10]. We describe for the first time a case of extraskeletal Ewing's sarcoma isolated in the maxillary sinus without involving the nose. The unusual site of this tumour makes this case apparently unique. However, as the ethmoid sinus and nose Ewing's sarcomas were named as an extraskeletal type in the literature, we decided to define this tumour as extraskeletal [3, 11]. The other factors such as tumour location in the maxiller sinus and the limited erosive area in the inferior orbital rim led us to believe that the tumour originated from the soft tissue. The cases of extraskeletal Ewing's sarcoma involving nasal cavity and/or paranasal sinuses are listed in Table 1. Differential histological diagnoses included malignant lymphoma, rhabdomyosarcoma, olfactory or secondary neuroblastoma [6]. A morphologically distinction between these entities may be difficult, particularly if biopsy material is limited. In the past, distinguishing Ewing's sarcoma from other small round neoplasms was difficult. But today immunohistochemistry and electron microscopy are of major importance in establishing a diagnosis in this group of neoplasms [6]. In our case, immunostaining of the tumour cells showed they were positive for CD99 which is truly definitive for Ewing's sarcoma. Neuroendocrine neoplasm's and the olfactory neuroblastoma show neurosecretory granules in the cells and react with relevant markers, such as neuron specific enolase (NSE), chromogranin and others [6]. Immunohistochemical staining for myoglobin and desmin are positive in rhabdomyosarcoma [6]. Immunohistochemical staining for LCA is positive in lymphoma [6]. In our case there was a slight reaction to NSE, but no reaction to LCA, desmin, CD20, CD30, and synaptophysin. The respective diagnoses of rhabdomyosarcoma, neuroblastoma and malignant lymphoma have been eliminated.

Soft tissue sarcomas differ from the usual bone-related Ewing's sarcoma in that they affect a younger population, usually in the second to fourth decade of life. Angervall and Enzinger [7] reported in 1975 that 60% of the patients die of disease within the first year after diagnosis, and at the time of their paper only 26% were alive without disease. Most of the recurrences were local in nature. They reported that metastatic disease typically went to the lungs or bone with only 7% going to lymph nodes [7].

It has been recommended to combine surgery, radiation and chemotherapy whenever possible for either bony or soft tissue related Ewing's sarcoma of the head and neck region [12]. Extraskeletal Ewing's sarcoma is a tumour sensitive to multimodality treatment. Early awareness and treatment of this rare disease and wide resection, followed by chemotherapy and radiotherapy might improve patient's long term survival. We recommended combined therapy to the patient, but she did not accept the surgical therapy. Therefore the patient was treated with chemotherapy and radiotherapy.

In summary, a case of extraskeletal Ewing's sarcoma in a 16-year-old female patient is the first described case of this tumour isolated from the maxillary sinus. Its treatment using chemo-radiotherapy is also presented. Early and accurate diagnosis, combined surgery, radiation and chemotherapy and extended follow-up are crucial due to this type of neoplasm.

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