Case Report / *Olgu Sunumu*

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Primary Ewing's Sarcoma of the Nasal Cavity: A Rare Case Report

Nazal Kavitenin Primer Ewing Sarkomu: Nadir Bir Olgu Sunumu

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Abstract

Primitive neuroectodermal tumor/Ewing's sarcoma (EWS) is a very rare tumor in the nasal cavity. It can be easily confused with sinusitis, polyps and other diseases. The most common symptoms are nasal congestion, epistaxis, olfactory disorders and facial pain. In our case, there was a huge tumor developed in the left nasal cavity and invaded orbita detected in physical examination. The diagnosis was EWS and the patient was given chemotherapy and radiotherapy. A rare nasal cavity tumor, EWS, is presented in this paper.

Keywords: Ewing's sarcoma, nasal cavity, sinonasal mass

Öz

Primitif nöroektodermal tümör/Ewing sarkomu (EWS) nazal kavitede çok nadir görülen bir tümördür. Nazal kavitede sinüzit, polip ve diğer hastalıklarla rahatlıkla karışabilirler. En sık rastlanan semptomları burun tıkanıklığı, epistaksis, koku alma bozukluğu ve yüz ağrısıdır. Olgumuzda sol nazal kaviteden kaynaklanan, orbitayı invaze eden dev tümör mevcuttu. EWS tanısı konulan hastaya kemoterapi ve radyoterapi uygulandı. Bu yazıda nadir bir nazal kavite tümörü olarak görülen EWS sunulmuştur.

Anahtar Sözcükler: Ewing sarkomu, nazal kavite, sinonazal kitle

Introduction

Sinonasal tumors constitute 0.2-0.8% of all body tumors and 3% of upper airway tumors. 77% of these tumors arise from the maxillary sinus (1). 80% of tumors of this area are squamous cell carcinoma, 10-15% are adenocarcinoma and adenoid cystic carcinoma. 4-6% are lymphoma, melanoma, esthesioneuroblastoma and sarcomas (1,2). In this article, diagnosis, definitive diagnosis and treatment attributes of rarely seen nasal cavity-derived primitive neuroectodermal tumor (PNET)/ Ewing's sarcoma (EWS) in a 21-year-old female patient are discussed in the light of the literature.

Case

A 21-year-old Syrian female patient was admitted with the complaints of nasal congestion and swelling pushing the left-eye, loss of vision, headache and facial pain (Figure 1). Endoscopic nasal examination revealed a tumoral mass with irregular margins and hyperemic appearance which was completely occupying the left nasal cavity. The mass was pushing the eyeball forward. There was also left eye ectropion and inability to close the eyelids completely. The mass was detected in the left maxillofacialregion. Her laboratory findings and medical history were unremarkable. Magnetic resonance imaging (MRI) demonstrated a mass filling the left periorbital area, ethmoid sinuses and reaching the maxillary bone and causing wide destructions. Since the mass had significant soft tissue components, the left maxillary sinus was observed to be obstructed and extradural-intracranial extension was seen in the left temporal lobe anteriorly. Left frontal and temporal lobes were pushed slightly by the mass. No calcification was observed in the mass (Figure 2). The biopsy result of the sample collected from the nasal mass was reported

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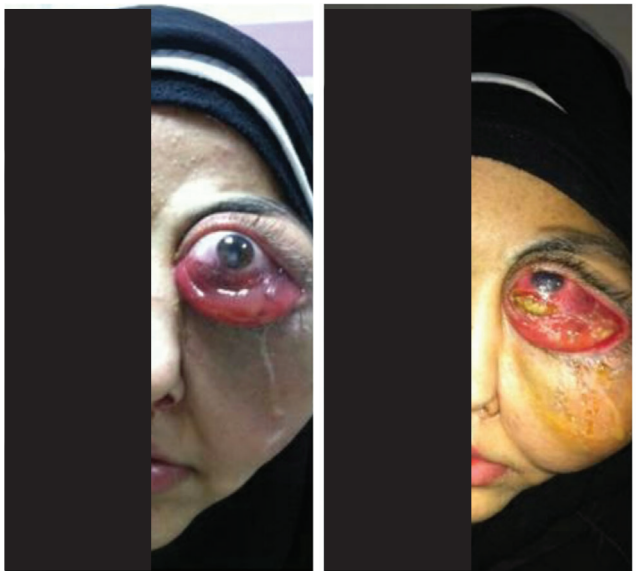

Figure 1. Extending tumor from left nasal cavity to orbita (left figure) and progression development after chemotherapy and radiotherapy (right figure)

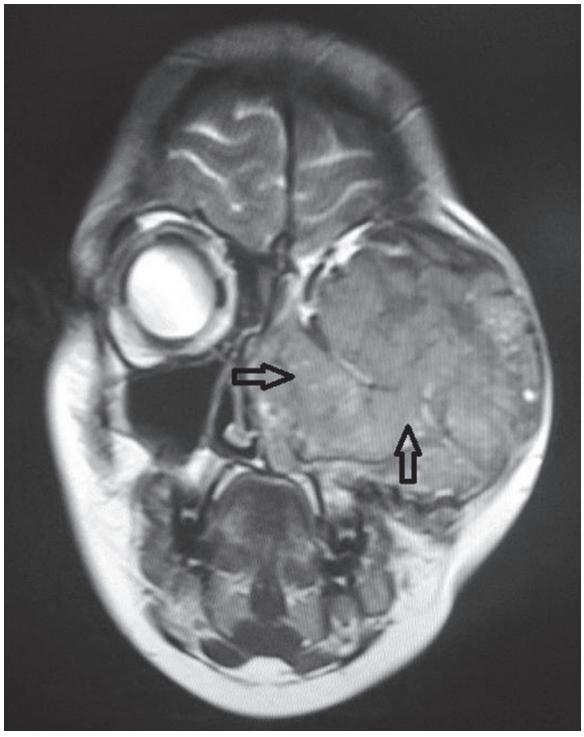

Figure 2. Magnetic resonance imaging of paranasal sinus

as PNET/EWS. Pathological examination revealed small round cells consisting of uniform hyperchromatic nucleus structures, nodular epitheloid cells and necrosis (Figure 3). Immunohistochemistry revealed positive CD99, vimentin and neurofilament staining specific to PNET. Pale positive staining with FLI-1 was detected. Negative staining with

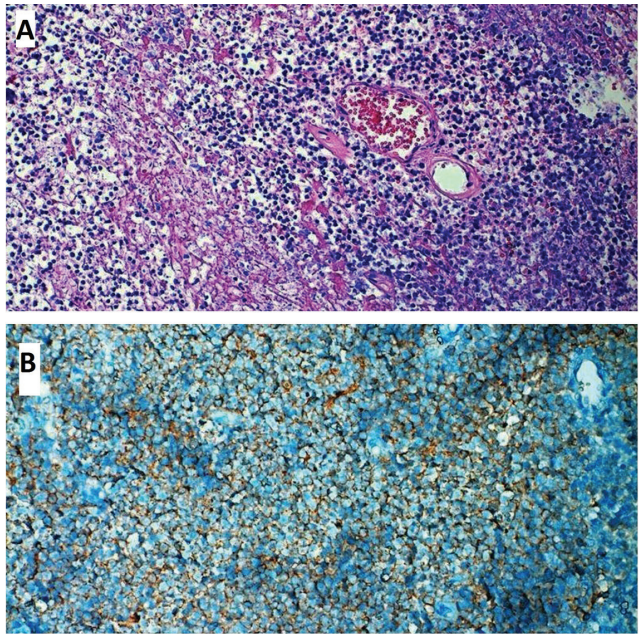

Figure 3. A) Cellular areas consisting of uniform small round cells, appearance of nodular epithelioid cells and necrosis (hematoxylin and eosin staining-superior picture), **B)** Characterization of membranous staining with CD99 (immunohistochemical staininginferior picture) (microscopic imaging 200 magnification)

CD45, lithocholic acids (LCA), pancreatin, neuron-specific enolase (NSE), terminal deoxynucleotidyl transferase, CD34, synaptophysin, desmine and chromogranin were observed (Figure 3). Lymphoma, neuroblastoma and carcinoma were eliminated since the samples were negative for CD45, LCA and NSE. Positron emission tomography (PET) is used for distant metastasis PET revealed a 77x60x57 mm hypermetabolic invasive mass involving the left maxillary sinus, left nasal cavity, left retro-orbital area, and left eyeball and destructing the bone structures, left rectus and pterygoid muscles. There was no distant metastasis. The mass was unresectable and chemotherapy was planned. The patient received treatment with vincristine (1.5 mg/m^2), mesna (1200 mg/m^2), cyclophosphamide (1200 mg/m^2), and doxorubicin (30 mg/m^2) (1st and 2nd day). Vincristine treatment was repeated in the 8th, 15th and 22nd days. Physical examination and MRI revealed chemotherapy-resistant progressive disease (Figure 1). The patient was given radiotherapy, however, she died during radiotherapy. The patient provided consent for publication. Informed consent was obtained from the patient.

Discussion

Tumors and lesions similar to tumors formed by cells having poorly differentiated or undifferentiated histologic pattern make the diagnosis difficult in the sinonasal area. Between the similar lesions, there are lymphoepithelioma

(undifferentiated carcinoma), retinoblastoma, olfactory neuroblastoma, medulloblastoma, small cell undifferentiated carcinoma, malign melanoma, lymphoma, plasmacytoma, rhabdomyosarcoma, synovial sarcoma and EWS. They should be differentiated for prognosis and treatment (3). Primary EWS of the head and neck region is an extremely rare condition accounting for only 4-9% of all EWS. 20-30% of patients with EWS patients have metastasis at the time of diagnosis. However, those with EWS of the head and neck region less frequently present with metastasis at the time diagnosis (4). Our patient had no metastasis. Rapidly progressing malignant diseases of the nasal and paranasal sinuses usually show orbital involvement. Sinonasal tumors often occur in patients between 50 and 70 years of age. It occurs 2 times more often in males. They are diagnosed in the advanced stage because of its rarity and symptoms mimicking the inflammatory diseases. Tumor shows symptoms when they are exceeded the sinus edge. The most common symptoms are nasal congestion, epistaxis and ache in the face. They can cause proptosis, diplopia and loss of sight by invading the orbit. If the tumor extends soft tissue by doing bone destruction, it can cause facial asymmetry as in the patient presented in this paper. Today, the disease can be diagnosed in earlier stages by endoscopic examination, computed tomography (CT) and MRI (1). Calderón-Garcidueñas et al. (2) reported that the time elapsed between the onset of the first symptom and diagnosis was 6-18 months in a series of 256 patients. Additionally, they found that only 1 of 4 tumor cases could be diagnosed when it was in the sinus. First symptoms were nasal congestion (63%), nasal bleeding (43%) and rhinorrhea in their series. Also, they reported pathological results as epidermoid carcinoma in 27%, non-Hodgkin lymphoma 24.5%, malign melanoma 14.4%, adenocarcinoma in 11% of patients (2). In sinus tumors, MRI is more suitable than CT for differentiating inflammatory disease from tumoral lesions. Therefore, MRI is the most important radiologic tool in the diagnosis of paranasal sinus tumors. PNET includes EWS, peripheral neuroepithelioma and poorly differentiated neoplasms including Askin tumor. PNET was first defined by Hart and Earle (5). These tumors are derived from malign small neoplasm cells, and even though generally seen in children, they can be seen in every age. EWS was first described by James Ewing firstly in 1921 as a malign tumor occurring in the shaft of long bones in children and young adults. While 47% of tumors develop in long bones, 19% in pelvis and 12% in the ribs, calvarial tumors are found in only 1-6% of cases (6). Tumor reaches to the soft tissue in 90% of patients. The tumor seen more frequently in males and there is no gender predominance for primary calvarial extension. The main clinical symptoms and findings are increased intracranial pressure, papilledema and headache depending on the intracranial extension of the tumor (7). Head and neck EWS rarely remains in the primary area and there are only a few reported cases of EWS of the nasal cavity (8). EWS is formed by uniform-looked undifferentiated small round tumor cells with thin fibrovascular septa separating into lobules or usually joined together in sheaths. Similarly, as it is in our case, it is usually vascular, its hemorrhagic areas and common necrosis areas are more frequent. Tumors neighbouring bone structures can cause osteosclerotic changes; it can progress with lytic lesions (9). EWS responds well to the radiotherapy, multi-agent chemotherapy and surgery. Treatment protocols should be planned individually for each patient. Chemo-radiotherapy combination after surgery is beneficial for prolonging survival (10). The most effective treatment is provided by radical surgical approach with pre-operative radiotherapy or post-operative radiotherapy in paranasal sinus cancer patients. In our case, the patient was not treated with chemotherapy combined with radiotherapy but the patient died. The 5-year survival rate is around 55-60%. Local recurrence is the leading cause of death (2). Sinonasal location of EWS is rare. In addition to pathologies, such as sinusitis and nasal polyps which encountered often in the sinonasal area, tumoral lesions should be considered. In this disease group, early diagnosis is very important. Apart from otorhinolaryngologists, ophthalmologist should have the knowledge about the symptoms and findings. Detailed head and neck examination including cranial nerves, endoscopic examination of the nasal cavity and paranasal sinuses and radiologic examinations should be done and treatment must be started immediately in all patients. In the postoperative period, the patients should be checked every month in the first year, every two months in the second year, and every four months in the third year.

Authorship Contributions

Surgical and Medical Practices: D.A., S.Ç., İ.Y. Concept: D.A., S.Ç., İ.Y. Design: D.A., S.Ç., İ.Y. Data Collection or Processing: D.A., K.K. Analysis or Interpretation: D.A., Ş.S.K. Literature Search: D.A. Writing: D.A.

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