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A Rare Case of Primitive Neuroectodermal Tumor of the Amygdala: A Case Report and Literature Review

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Abstract

Background: The primitive neuroectodermal tumor (PNET) belongs to the family of malignant small round cell tumors with neuroepithelial differentiation. Isolated cases of PNET have been reported in visceral sites, such as the kidneys, uterus, ovaries, testicles, bladder, and pancreas. The occurrence of these tumors in the head and neck region is very rare. **Purpose:** We report the case of a 37-year-old patient with a PNET of the amygdala and a favorable outcome after induction chemotherapy followed by exclusive radiotherapy. **Results:** The patient exhibited a significant clinical and radiological response to the first-line chemotherapy regimen (VAC-IE) followed by radiotherapy. The treatment led to a complete clinical and radiological response. Currently, the patient is alive and in good health, with no signs of local recurrence or metastasis three years post-treatment. **Conclusion:** PNETs of the amygdala are extremely rare and aggressive. A combined therapeutic approach involving surgery, chemotherapy, and radiotherapy is essential for effective treatment. This case demonstrates that aggressive treatment can lead to favorable outcomes even in rare tumor locations.

Subject Areas

Neurology, Oncology

Keywords

Primitive Neuroectodermal Tumor (PNET), Amygdala, Chemotherapy, Radiotherapy, Surgery

1. Introduction

The primitive neuroectodermal tumor (PNET) is a term used to describe a group of highly malignant small round cell tumors of neuroectodermal origin with variable cellular differentiation [1]. Although PNET primarily occurs in the central nervous system, peripheral PNETs have also been reported. Isolated cases of PNET have been documented in visceral sites, such as the kidneys, uterus, ovaries, testicles, bladder, and pancreas [2] [3]. They rarely arise in the head and neck region, with only five cases reported in a series of 118 patients including the parotid gland [4] [5]. Management of PNET necessitates aggressive surgical treatment of local disease with chemotherapy and radiotherapy to treat residual disease and prevent metastatic or recurrent disease. We present a case report of a rare primary PNET of the amygdala. To our knowledge, fewer than 10 cases of PNET in the head and neck region have been reported in the literature.

2. Patient and Observation

We report the case of a 37-year-old female patient with no notable medical history. Six months prior to diagnosis, she presented with a gradually increasing right-sided oral swelling accompanied by a deterioration in general condition, prompting her to seek consultation with an otolaryngologist.

The clinical examination under fiberoptic endoscopy revealed a right lateral oral mass involving the oropharynx. A biopsy under endoscopic guidance was performed, which returned negative for signs of malignancy.

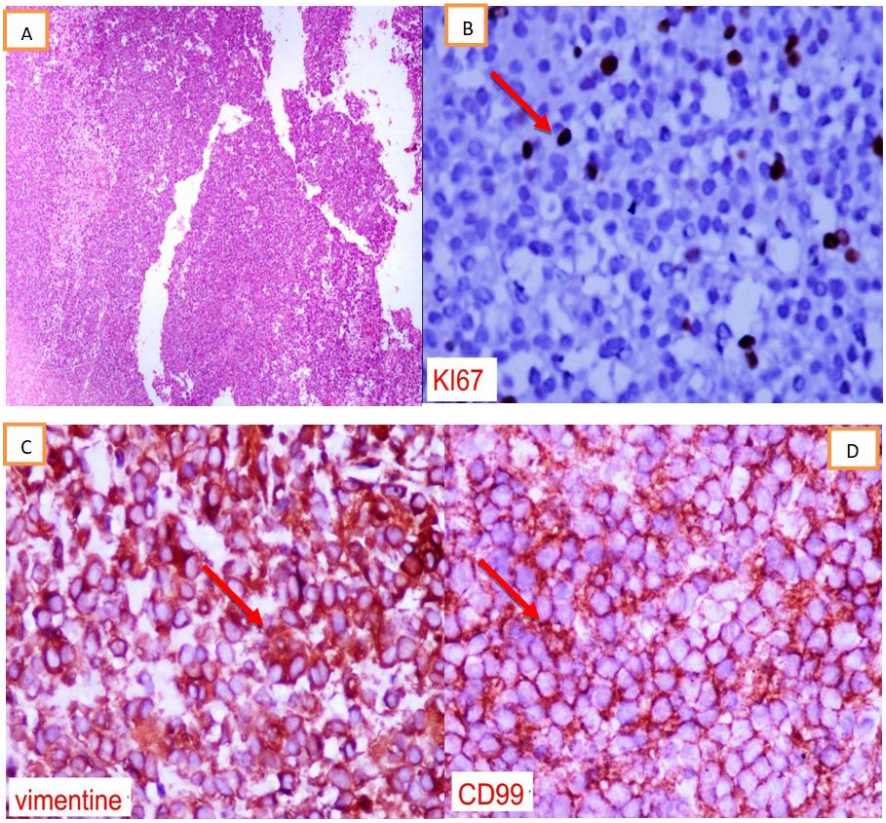
Cervicofacial MRI imaging was conducted, revealing a right amygdalartumoral process protruding into the pharynx with hyperintense T2 signal, measuring 47 × 40 × 50 mm. This tumor exhibits focal areas of necrosis. It completely occupies the parapharyngeal space and invades the upper palate veil. Filling of the parapharyngeal and retro-molar space is noted externally. Right submaxillary lymph nodes measuring 16 × 9 mm were observed.

Due to the worsening symptoms, including the onset of inspiratory dyspnea, the patient underwent a tracheotomy and R2 resection of the tumor mass, involving excision by fragmentation of the obstructive tumor from the oral cavity, aimed at debulking for the purpose of alleviating obstruction.

An anatomopathological study of the operative specimen revealed an undifferentiated tumor proliferation consisting of small round cells without lobulated structures, characteristic of a PNET. A comprehensive immunohistochemical panel was conducted. The neoplastic cells showed diffuse immunoreactivity for vimentin, while epithelial markers (cytokeratin) were negative. There was strong positivity for the CD99 marker, while other markers including CD20, CD34, CD56, PS 100, and chromogranin A were all negative (**Figure 1**).

A CT scan of the chest, abdomen, and pelvis for staging was performed, revealing no evidence of distant metastasis.

The patient underwent first-line chemotherapy with VAC-IE regimen, showing a very good clinical and radiological response. The evaluation CT scan after



Note: (A) Microscopic appearance of amygdala PNET; (B) Nuclear labeling by KI67; (C) Cytoplasmic labeling of tumor cells by vimentin; (D) Membranous labeling of tumor cells by CD99.

Figure 1. Histological characteristics of amygdala PNET.

3 cycles demonstrated a significant decrease in size and extent of oropharyngeal thickening (15 mm vs. 45 mm). She received an additional 3 cycles of chemotherapy.

Subsequently, following a multidisciplinary discussion, the patient received radiotherapy for residual tumor, delivering 70 Gy over 7 weeks, resulting in complete clinical and radiological response.

Currently, the patient remains alive with good local-regional control and general condition, 3 years after completion of treatment, without any local disease recurrence or signs of metastasis.

3. Discussion

Primitive neuroectodermal tumors (PNETs) are rare tumors, estimated to represent 1% of all sarcomas. They are frequently described in children and young adults, mainly in sites such as the buttocks and thighs, followed by the shoulder and arm [1]. Most published series reveal a rarety of PNET cases in the head and neck region [6]-[8]. The oral cavity, nasal cavity, and skull are distinct sites in the head and neck region where PNETs have been reported [9].

In the head and neck region, PNETs have been described in the maxilla, orbit,

and soft tissues of the neck. However, to our knowledge, there is only one reported case of PNET in the parotid gland in the literature [1]. In the head and neck region, clinical symptoms vary depending on the site of involvement but invariably include pain and swelling of surrounding structures due to mass effect (our experience).

Primitive neuroectodermal tumors (PNETs) of the amygdala (as in our case) typically present as a rapidly growing, painful or painless mass compressing surrounding structures. On high-resolution computed tomography, they appear as irregular masses with heterogeneity and hypodensity, with contrast enhancement. On magnetic resonance imaging (MRI), the tumor is isointense compared to muscle on T1-weighted images, while it is hyperintense on T2-weighted images [10].

Microscopically, PNETs are challenging to distinguish from other round cell tumors; they consist of small, uniform round or oval cells containing cytoplasmic glycogen and are sometimes arranged around a central space filled with fibrillar extensions of the cells [10].

The immunohistochemical profile allows the pathologist to distinguish PNETs from other round cell tumors. PNETs are positive for MIC-2, an antigen derived from the MIC-2 gene, vimentin, S-100, specific neuronal enolase, desmin, CD 75, and neurofilament protein. Electron microscopy and immunostaining play a crucial role in differentiating PNETs from other tumors with a similar appearance, such as non-Hodgkin's lymphoma, neuroblastoma, rhabdomyosarcoma, etc. [9].

Primitive neuroectodermal tumors (PNETs) are extremely aggressive tumors that commonly metastasize to the lungs, bones, and bone marrow [11]. Micrometastatic disease is demonstrated in the bone marrow using reverse transcription polymerase chain reaction (RT-PCR) technology in up to 30% of patients considered to have localized disease. In a large series of 54 patients, the metastasis rate ranges from 20 to 31%, with a survival rate of less than 25% [10].

Surgery with clear margins combined with chemotherapy and radiotherapy is the treatment of choice for PNET tumors. However, complete resection of PNETs in the head and neck may be challenging due to involvement of vital structures. Chemotherapy and radiotherapy remain a viable alternative when surgery is difficult [10]. In the case reported here, our patient is in good general condition with no recurrence 3 years after treatment. The adopted protocol consisted of 6 cycles of VAC-IE (dose) followed by radiotherapy at a dose of 70 Gy delivered over 7 weeks.

4. Conclusion

Primitive neuroectodermal tumors (PNETS) are rare malignant small round cell tumors, particularly in the head and neck region. This case report presents a unique instance of PNET arising in the amygdala, adding to the limited existing literature. The patient's favorable outcome following aggressive multimodal treat-

ment highlights the importance of a comprehensive approach combining surgery, chemotherapy, and radiotherapy.

Ethical Considerations

This study was conducted in accordance with the ethical principles of the Declaration of Helsinki.

Conflict of interest

The authors declare no conflicts of interest.

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