

Primitive neuroectodermal tumor in advanced age: case report

Tumor neuroectodérmico primitivo em idade avançada: relato de caso

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ABSTRACT

Objectives: Primitive neuroectodermal tumors (PNETs) are aggressive neuroectodermal neoplasms and are originated from neuroectodermal tissues. They belong to a larger group of tumors known as Ewing tumors and are unusual in adult population. This case reports PNET in the oldest patient quoted in literature and established it as a differential diagnose to soft tissue tumors in advanced age. **Case report:** Female, 69 years, with swelling and pain in the nasal cavity. Images revealed expansive mass with soft tissues density occupying paranasal sinuses, nasopharynx and rhinopharynx diminishing the air column. Histopathologic findings were compatible with PNET. **Conclusion:** Primitive neuroectodermal tumors are more often found in children. This current case report call attention to a 69-years-old woman with a PNET on nasopharynx. Once the patients' age and the disease site are both extremely unusual, this case report seeks to contribute to the diagnosis of rare PNETs presentation forms.

Keywords: Sarcoma, Ewing; Neuroectodermal tumors, Primitive; Soft tissue neoplasms.

RESUMO

Objetivos: Os tumores neuroectodérmicos primitivos (PNETs) são neoplasias neuroectodérmicas agressivas e são originários de tecidos neuroectodérmicos. Eles pertencem a um grupo maior de tumores conhecidos como tumores de Ewing e são incomuns na população adulta. Este caso relata PNET em paciente mais velho citado na literatura e o estabelece como um diagnóstico diferencial para tumores de tecidos moles em idade avançada. **Relato de caso:** Feminino, 69 anos, com edema e dor na cavidade nasal. As imagens revelaram massa expansiva com densidade de tecidos moles ocupando seios paranasais, nasofaringe e rinofaringe diminuindo a coluna de ar. Os achados histopatológicos eram compatíveis com o PNET. **Conclusão:** Os tumores neuroectodérmicos primitivos são mais frequentemente encontrados em crianças. Este relato de caso atual chama a atenção para uma mulher de 69 anos com PNET na nasofaringe. Uma vez que a idade do paciente e o local da doença são extremamente incomuns, este relato de caso procura contribuir para o diagnóstico de formas raras de apresentação de PNETs.

Descritores: Sarcoma, Ewing; Tumores neuroectodérmicos primitivos; Neoplasias de tecidos moles.

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INTRODUCTION

Ewing sarcoma family of tumors are rare malignancies derived from a common cell of origin.¹ These neoplasms normally arise from bone or soft tissue, and are histologically characterized by small round blue cells. Regarding their molecular basis, the Ewing Family of tumors share a pattern of non-random chromosomal translocations. About 90% of cases occur due to translocation of chromosome 11 and 22, t(11,22)(q24;q12).²

PNET (primitive neuroectodermal tumor) first called peripheral neuroepithelioma and Askin's tumor of the chest wall, is a more differentiated member of this tumor family. It was first described in 1918 by Stout as a tumor of the ulnar nerve, comprising characteristics of a sarcoma, but composed of small round cells arranged as rosettes.³

PNET can present with a wide range of clinical features depending on the affected site, though pain and swelling of the surrounding structures are the more common signs and symptoms.⁴

It is recognized as an aggressive tumor, with an accelerated progression and high relapse rate, requiring a multiprofessional approach in term soft treatment.⁵ Even though fewer than 25% of patients have overt metastases at the time of diagnosis, it is considered a systemic disease, given the high incidence of subclinical metastatic disease.⁶

PNET occurs more frequently in individuals aged 10 to 20 years, thus being considered as child and adolescent neoplasm.² Its prevalence is slightly higher in males.⁷

Disease occurrence in older patients is considered an adverse prognostic factor, although it is not known if this is a consequence of the biological differences or differences in treatment approach. There is some evidence that older patients treated the same way as the younger ones may have comparable survival outcomes^{8,9}. Here we report a PNET case in an extremely unusual age of on set forth is neoplasm.

CASE REPORT

A 69-year-old female presented with complains of swelling and pain in the nasal cavity. She was submitted to Nasal Sinus's CT (Computed Tomography) about 6 months after symptoms onset. Images revealed an expansive mass with soft tissues density occupying completely the maxillary, frontal, ethmoidal and left sphenoidal sinuses, as well as the nasal cavity and fossa, also extending itself to the left side coana and rhinopharynx, diminishing the air column. There was no cleavage plane with the left nasal turbinates, though it was clear that the adjacent osseous areas were remodeled and thinned including the maxillary sinus's posterior wall (Figure 1).

It was also evidenced an infundibular enlargement of the Ostium meatal complex and bulging of the papyracea blade on the left side. CT scan indicated calcification are as associated as well, which brought, at first, the hypothesis of a nasal sinus polyposis (Figure 2).

Three months later, an endoscopic nasal exploration with biopsy – sinusectomy with Caldwell Luc technic – was performed. The anatomopathological exam revealed a Large Cells Malignant Undifferentiated Neoplasm on the sinus mucosa with positive border, and the immunohistochemistry study demonstrated a primitive neuroectodermal tumor pattern with cell proliferation index of 50-55%.

At the time of diagnosis, the patient had a decent performance status (ECOG 1) and no significant comorbidities, being a candidate for aggressive treatment. Patient's pain was controlled with oral morphine and systemic chemotherapy was initiated. Patient was prescribed doxorubicin plus cyclophosphamide plus vincristine (VAC) on odd cycles, alternating with iphosphamide plus etoposide on even cycles (VAC/IE regimen).¹⁰

Unfortunately, after being submitted to 2 cycles of chemotherapy, the patient died from a septic shock.

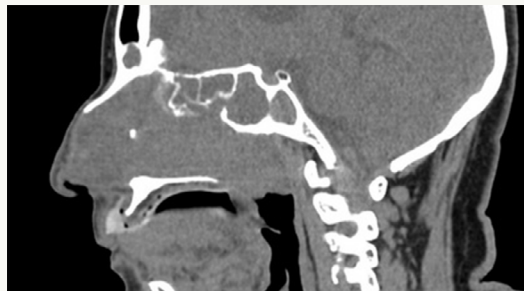


Figure 1. Expansive lesion with soft tissue density and permeation calcifications occupying all paranasal sinuses on the left, besides the fossa and nasal cavity, with extension to the coana and rhinopharynx.



Figure 2. Determines bone remodeling with adjacent erosion, infundibular widening of the ostiomeatal complex and bulging of the papyracea blade on the left.

DISCUSSION

To our knowledge, this is the oldest reported patient reported with PNET in medical literature. PNETs are usually classified as a child or adolescent disease, but as shown, it may occur in older patients as well.¹¹

PNET diagnosis is performed through histological analysis,⁷ with immunohistochemistry support if necessary. Testing for the EWS translocation may be necessary for diagnosis confirmation.² In the present case, diagnosis was made without genetic testing due to highly suggestive histological analysis lack of material for EWS translocation testing.

Most guidelines for PNETs treatment come from data on children and adolescents. Local control provides a survival benefit for non-metastatic disease, but all patients should receive systemic chemotherapy¹².

The VAC/IE regiment has shown superior overall survival for non-metastatic patients and superior progression-free survival for all patients when compared to VAC alone.¹⁰ VAC/IE has since become the standard treatment for non-metastatic PNETs. It should be noticed, however, that only 13% patients included in that trial were older than 18 years.

Lack of consistent evidence, poor performance, and worse treatment tolerance may account for a less favorable prognosis when compared to the same disease in younger patients. However, there is some evidence that this disease is still chemosensitive in these patients. A recent retrospective study¹³ found a 70% three-year disease-free survival rate for patients older than 19. Most patients in this study were treated with the VAC/IE regimen.

CONCLUSION

PNET tumors are typically a children and adolescent disease, but as our report shows, it can also appear in older populations - such as this 69-year-old woman.

Thus, PNET should not be ruled out as a differential diagnosis in older patients based on age alone. Treating such patients with PNETs is a clinical challenge, but aggressive chemotherapy should not be denied to them.

AUTHOR'S CONTRIBUTION

Juliana de Almeida Zavarize: Collection and assembly of data, Conception and design, Manuscript writing.

Octávio Sakahara Saito: Collection and assembly of data, Manuscript writing.

Fernanda Proa Ferreira: Manuscript writing, Provision of study materials or patient.

Frederico Leal: Data analysis and interpretation, Final approval of manuscript, Manuscript writing.

Denise Junqueira Maia Soares: Collection and assembly of data.

Juliano Manzoli Marques Luiz: Collection and assembly of data, Manuscript writing.

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