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Primitive Neuroectodermal Tumors. State of Art

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INTRODUCTION

PNETs, also known as primitive neuroectodermal tumors, are extremely uncommon sarcomas that originate from the neural crest. It has been observed that the yearly incidence of these instances is 2.9 cases per million people between the ages of birth and 20.

Typically, these tumors originate from the bone and soft tissues, and they are more frequently discovered in individuals who are younger. The histological appearance of PNETs is similar to that of tiny, spherical cells that stain darkly and have varied differentiation. This is seen when the cells are examined using light microscopy. In addition, PNET tumors frequently exhibit aggressive behavior and are indicative of a bad prognosis. It is the existence of metastatic disease that is the most significant determinant in determining the prognosis. Due to the fact that the majority of patients present with metastases at the time of diagnosis, it is unfortunate that no pathognomonic symptoms have been discovered.

PNET is a condition that can be difficult to diagnose because of its varied histology and wide variety of anatomic origins. The diagnosis of PNET requires a mix of techniques, including immunohistochemistry, light or electron microscopy, and a comprehensive history. PNETs have traditionally been classified into two primary categories, namely central (cPNET) and peripheral (pPNET), respectively, according to the brain tissue from which they originate.8 pPNETs originate from peripheral nerves or soft tissue and are traditionally thought to be a member of the wider Ewing sarcoma (ES) family of cancers due to the chromosomal and histopathologic similarities between the two types of tumors.As an illustration, pPNET and ES often exhibit a chromosomal translocation that follows the pattern t(11;22) (q24;q12). pPNETs originate most typically in men in their second decade of life, notably among individuals of White and Hispanic ethnicity. cPNETs constitute a more varied range of tumors emerging from the central nervous system (CNS). Medulloblastomas, pinealomas, cerebral neuroblastomas, and primary CNS rhabdomyosarcomas have historically been included in this category of embryonal sarcomas that are situated in the central region of the embryo. Although the histology of medulloblastoma is substantially identical to that of cPNET, it is currently regarded to be physiologically separate from cPNET. Medulloblastoma is the most prevalent kind of central central brain tumor.111 Even if the frequency of cPNET is larger than that of pPNET, the male gender continues to be the more prevalent.

Even though they have certain similarities, pPNET and cPNET are two distinct disease processes that are distinct from one another in terms of their clinicopathologic features, therapy, and results. As a result of the rarity of PNET, healthcare practitioners have a limited amount of information to use in order to ensure that patients with both pPNET and cPNET are managed successfully.

key aspects of primitive neuroectodermal tumors (PNETs):

Aspect	Details
Definition	A group of highly malignant tumors composed of undifferentiated or poorly differentiated neuroectodermal cells.
Common Locations	Brain (CNS-PNET), peripheral nerves (Peripheral PNET), and other tissues such as bones and soft tissues (Ewing sarcoma family of tumors).
Histology	Small round blue cells, rosette formation, high mitotic rate.
Symptoms	Varies by location but may include headaches, seizures, focal neurological deficits, pain, swelling.
Diagnosis	MRI/CT scans, biopsy, histopathological examination, immunohistochemistry, genetic testing.
Treatment	Surgery, radiation therapy, chemotherapy, stem cell transplant.
Prognosis	Varies widely depending on location, size, metastasis, and response to treatment.
Common Genetic Alterations	EWSR1-FLI1 fusion gene (Ewing sarcoma), amplification of MYC family genes (CNS-PNET).
Associated Conditions	Li-Fraumeni syndrome, neurofibromatosis type 1.

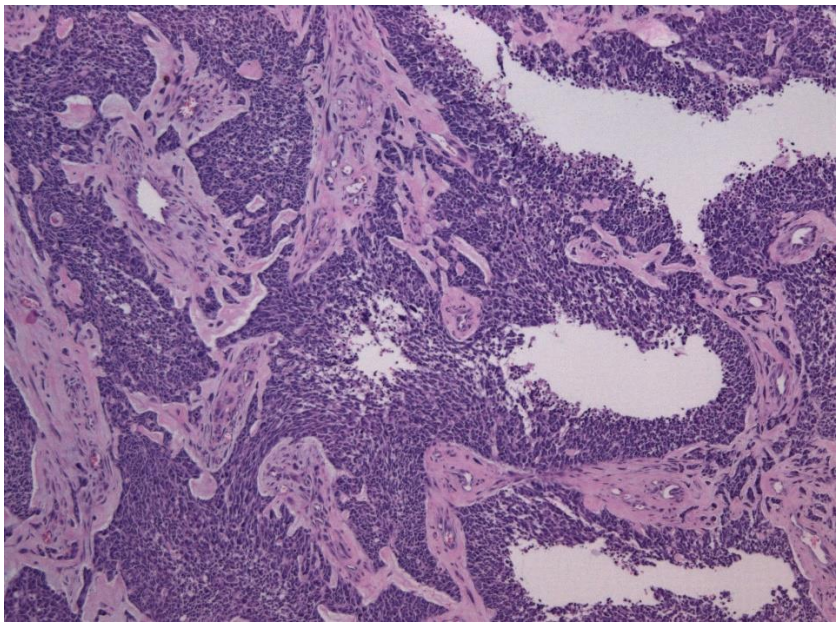

Figure 1. Glioblastoma PTEN component

DISCUSSION

Our analysis of NCDB is the most comprehensive examination of PNET based on demographic data that has been conducted to this point. When it comes to prior knowledge of PNET diagnosis, the majority of the information comes from tiny case series and studies. An evaluation that is based on the more general categories of cPNET vs pPNET has made it easier for us to compare aspects such as incidence, treatment patterns, and results. This is despite the fact that we acknowledge that every single diagnosis of PNET is distinct. Prior investigations have shown that White males appear to be at a higher risk for PNET diagnosis, and our data are consistent with those conclusions. Both the pPNET and the cPNET have demonstrated this to be the case. Males made up roughly 70 percent of the cases in a retrospective case study that included 36 individuals diagnosed with pPNET.a 13 The Surveillance, Epidemiology, and End Results (SEER) program at the National Cancer Institute (NCI) was examined for ES and pPNET for the most current time. Eighty-five percent of the 630 patients who were diagnosed with pPNET were white, and 54.8% of them were male. Additionally, data from the NCI SEER have been provided for diagnosis of cPNET and medulloblastoma spanning the years 1973 to 1998. In this particular instance, the scientists discovered that these central tumors were more prevalent in white people (82%) and males (63%). It is known that PNET diagnoses are more prevalent in younger populations; nevertheless, the average age of diagnosis in this cohort was somewhat older, coming in at 41.8 years. This may be explained by the heterogeneity of this data set as well as the fact that only patients who were older than 18 years old were reported. Providers should maintain a high level of suspicion regarding PNET diagnosis in this patient population, regardless of the patient's age.

The yearly incidence of PNET cases has remained steady throughout time, as seen by these data, which confirm this assertion. Furthermore, despite the fact that both cPNET and pPNET continue to be extremely uncommon, our findings of 125 instances per year are consistent with the figures that have been reported by other studies. In a pediatric population, Esiashvili et al. discovered that over the course of three decades, there were an average of 2.93 occurrences of ES/PNET diagnoses per million persons occurring year. In a similar vein, the incidence of ES and other soft tissue sarcomas was 2.95 per million, and the average percent change remained steady in another study that focused on cancer in children and adolescents.The incidence history of cPNET diagnoses is more varied than those of other diagnoses; nevertheless, more recent data imply that these patterns of diagnosis are stable as well. According to the findings of an analysis of two cancer registries, there was no discernible pattern in the prevalence of cPNET diagnoses among adolescents and young adults in northern Europe between the years 1990 and 2013. However, it was discovered that the number of specific diagnoses of medulloblastoma was decreasing. An intriguing finding was made by McNeil et al., who discovered that the number of diagnoses of PNET/medulloblastoma in the United States increased by 23% between the years 1970 and 1990. In recent years, immunohistochemistry has made significant advancements in the diagnosis of PNET, which is the best explanation for this disparity. The incidence of medulloblastoma has not altered, according to the findings of a review of the Central Brain Tumor Registry of the United States, which made a clear distinction between medulloblastoma and cPNET. On the other hand, when PNET diagnoses were included, the incidence increased between the years 1985 and 2002. Future research should modify its approach to determining the incidence of pPNET and cPNET, making careful to differentiate between distinct tumors (such as medulloblastoma) as a separate entity. This is because our understanding of these tumors is expanding beyond the realm of histology and into the realm of cellular biology.

PNET can arise from odd areas, which is something that surgeons and other medical professionals who treat pPNET need to keep in mind. It has been described that pPNETs may be found in the adrenal gland, the dura mater, and the small bowel mesentery. An other type of tumor that has been recorded is one that originates from the head and neck, namely the paranasal sinuses and the nasal cavity. Due to the fact that these tumors are extremely uncommon in all areas, finding the most effective treatment for them continues to be difficult. In many cases, a multidisciplinary strategy that includes chemotherapy, radiation therapy, and surgery is required. There is a growing corpus of research that supports aggressive local control, with the primary focus being on obtaining negative resection margins in both cPNET and pPNET. Surgeons who treat these tumors have the primary goal of removing all macroscopic disease while still preserving the most amount of neurologic and functional recovery possible.

In line with this guideline, seventy-seven percent of the patients in this cohort got surgical excision of the original tumor that they were experiencing. As a result of the fact that patients with pPNET are more likely to present with metastatic illness at the time of diagnosis than patients with cPNET, it is possible that resection is more straightforward for cPNETs than it is for pPNETs. Also, when essential structures are affected, it is probable that surgical resection will not be viable for these individuals at the beginning of the process. It is recommended that systemic chemotherapy be started initially in instances of ES/PNET because the majority of these tumors are amenable to different types of treatment. By doing so, a safer surgical approach may be used, hence reducing the likelihood of the tumor spreading.

Neoadjuvant chemotherapy is often administered in the form of a combination regimen, which may include vincristine, ifosfamide, doxorubicin, and actinomycin. The utilization of this chemotherapy method improves the capability of achieving full tumor excision with negative microscopic margins. Patients diagnosed with ES/PNET may be able to avoid receiving radiation from an external beam as a result of this. It should come as no surprise that a diagnosis of cPNET is independently related with an increased chance of obtaining radiation therapy in comparison to pPNET in individuals who have been diagnosed with personal network tumors. In point of fact, cPNETs are classified as radiosensitive tumors, and postoperative irradiation of the craniospinal axis has demonstrated a considerable survival advantage. When it comes to both forms of PNET, multimodal therapy is suggested, and before moving forward with any intervention, doctors need to give serious consideration to each option. There are a number of restrictions that should be taken into consideration with regard to the current investigation.

To begin, despite the fact that the NCDB is one of the biggest cancer registries in the world, it does not gather sufficient data on a number of crucial outcomes. For instance, the National Cancer Database (NCDB) does not include patterns of progression-free survival or progression-free recurrence of PNET, and as a result, these patterns are not included in this analysis. In addition, although the National Cancer Database (NCDB) normally includes information on TNM staging, the majority of these tumors do not have these data. This substantially restricts our capacity to examine outcomes for patients who have localized disease as opposed to metastatic illness. In addition, certain therapeutic regimens for chemotherapy and radiation are not provided. Next, there were data points that were missing, which hindered our ability to comprehend and report on the surgical strategy that surgeons who were caring for these patients took. Improvements in documenting and description of resections will make it possible to have a comprehensive grasp of initial treatment approaches. In conclusion, we acknowledge that the statistical significance that was discovered in this study would not be obtained in clinical settings due to the significant deviation in sample size that exists between aggregate data and treatment facilities. Despite these limitations, our research has shed light on significant patterns in epidemiology, treatment patterns, and outcomes for surgeons and physicians who are responsible for the care of patients with uncommon sarcoma subtypes.

CONCLUSION

PNET tumors are a kind of cancer that is uncommon but aggressive. They can develop in a variety of sites, but they have a preference for the soft tissues of young men who also identify as White. Multimodal therapy, which often includes radiation therapy, chemotherapy, and surgery, is typically required for treatment. Treatment with radiation is administered to patients with cPNET more frequently than it is to people with pPNET. cPNET is likewise more frequently susceptible to resection; yet, it is associated with a lower death rate at 90 days, although the total survival rate is comparable to that of pPNET. It is possible that future research that aims to understand the biology of PNETs and uncover distinctions between these two forms of tumors could assist improve therapy as well as outcomes for people who have these sorts of cancers who are undergoing treatment.

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