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Case report

Primary Primitive Neuroectodermal Tumor of the Parotid: An Unsuspected Diagnosis

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Abstract:

Primitive neuroectodermal tumors (PNETs) are extremely rare tumors. These neoplasms can occur at sites outside of the central nervous system. The head and neck location is unsual. We report a case with primitive neuroectodermal tumor (PNET) of the parotid. We analyze through this observation the clinical, histological and therapeutic characteristics of this entity.

Keywords: primitive neuroectodermal tumor; parotid; surgery; chemotherapy; radiation therapy

Introduction:

Primitive neuroectodermal tumors (PNETs) are malignant tumors of the central nervous system. These malignancies are usually found in children and young adults. The diagnosis of PNETs is based on light microscopy following identification of a small round cell tumor and on the immunohistochemical staining results. They can occur at sites outside of the central nervous system usually within bone, pelvis, the chest wall and rarely in the head and neck [1]. We present here an extremely rare case of PNET of the parotid gland.

Case report:

A-33-year-old woman presented with a recurrent rapidly growing mass in the left parotid of 12 months duration associated with *facial asymmetry*. She had an excision of this mass in another institution since 6 months. The initial histological examination showed a benign *mixed tumor of the salivary glands*. On physical examination, she had a facial paresis. A left submandibular mass was noted. The tumor size was 6 cm. Upon clinical examination, a painless, non swelling and fixed mass (4x4cm) was found on the left parotid. There was no nodal involvement. *Magnetic resonance imaging* (*MRI*) of head-neck showed a soft-tissue mass of heterogenous density in the left side of the parotid gland (Figure 1). The size of the tumor was measured 31*35*42mm. The lesion appeared heterogeneous hypointense on T1 weighted images and hyperintense on T2 weighted images. The tumor had irregular contours with invasion of *pterygoid and masseter* muscles. *She underwent* surgical excision with total *parotidectomy*, including *facial nerve sacrifice*. Histological examination showed a polylobulated tumor with peripheral hemorrhagic reshuffle. *Tumor* cell *proliferation infiltrating parotid* gland *parenchyma with nodular* or *diffuse architecture was observed*. Solid sheets of *small-to medium*-sized cells with round to oval nuclei, basophilic cytoplasm and mitotic figures were revealed (Figure 2). *There were no lymph node* metastases.

The immunohistochemistry showed positivity for cytokeratin, CD99 and negativity for S-100 protein, CD45, synaptophysin, chromogranin, CD56, vimentin, epithelial membrane antigen(EMA) and CD34. On the basis of these findings, the lesion was confirmed to be a pPNET of the parotid gland. Cytogenetic analysis showed the specific translocation involving the EWSR1 gene on chromosome 22q12. This specific genetic abnormality was demonstrated by FISH (Figure 3). At the *time* of this *writing*, our patient is alive. She started start adjuvant radiation therapy and chemotherapy (VDC/IE regimen).She received 9 courses of chemotherapy. She finished the treatment one month ago. The patient has been under close observation since the treatment and there have been no signs of recurrence.

Discussion:

We describe a case of localized primary primitive neuroectodermal tumor (PNET) of the parotid in a 33 year –old man. PNET are rare tumors that belong to the Ewing's sarcoma family. They represent 1% of all sarcomas [2]. Commonly, PNET is seen in children and young adults. It has a slight *predilection for males* and occurs in bone and within soft tissues [3]. PNETs are rarely noted in the head and neck region. They can be found anywhere within the body, particularly in the trunk and extremities. Cases of PNET occurring in the genital tract, chest wall and retroperitoneal cavity have been reported [4]. In the head and neck region, PNET is usually found in the jaw, mandible and maxilla [5] . The clinical presentation is often associated with pain, swelling and cranial nerve deficits [6]. The patient in the current case presented with rapidly growing mass in the left parotid associated with *facial asymmetry*. PNET of the parotid is extremely rare. Only 6 cases have been reported in the literature (table 1). The imaging features of pPNETs are non-specific but are essential for diagnosis and surgical planning. On MRI, the tumor appears isodense or slightly hypodense on CT scan. Cystic or necrotic area with non-calcified masses are usually observed. On MRI, [7]. Histologicallay, the most helpful diagnostic feature is the presence of rosettes, usually of Homer-Wright type[6]. However, it is usually difficult to differenciate PNETs from other small roud-cells cancers. Immunohistochemically is useful for the differential diagnosis. In fact, PNETs consistently stain immunohistochemically for vimentin and

CD99. Tumor cells are negative for desmin and cytokeratin. Molecular and cytogenetic studies are essential to confirm the diagnosis. A chromosomal translocation t (11;22)(q24;q12) is found in about 90% of PNETs [6].

The cornerstone of multidisciplinary treatment is surgery followed by radiotherapy with doses

ranging from 45 to 60 Gy and chemotherapy. Our patient had surgery followed by chemotherapy(VDC/IE regimen) and radiation therapy. Surgical excision with negative margins is paramount for local control. But, this surgery can be difficult in the head and neck region in cases of involvement of vital structures. Radiation therapy and chemotherapy can be considered in these cases [8]. The most effective chemotherapeutic regimen for PNETs are Vincristine, Cyclophosphamide, Doxorubicin ,Iphosphamide and Etoposide. Despite the various treatments available, the prognosis of PNETs remains poor.

Close cooperation among surgeons, radiation and medical oncologists is needed for the management of PNET of the head and neck.

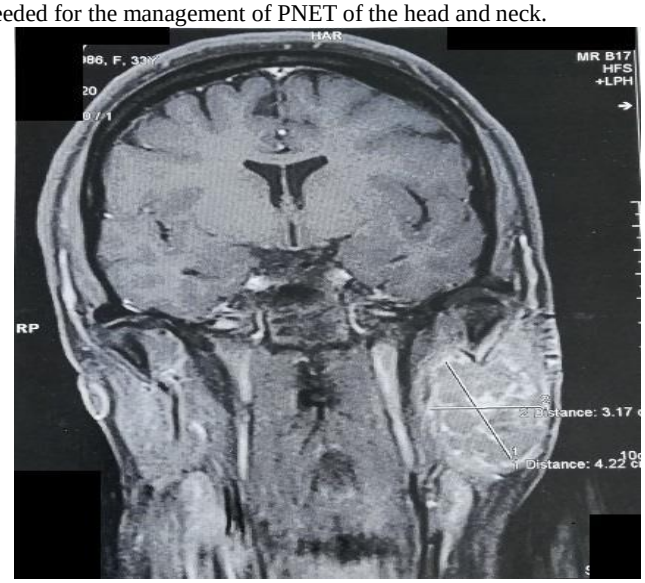

Figure 1: *Magnetic resonance imaging* (*MRI*) of head-neck showed a soft-tissue mass of heterogenous density in the left side of the parotid gland

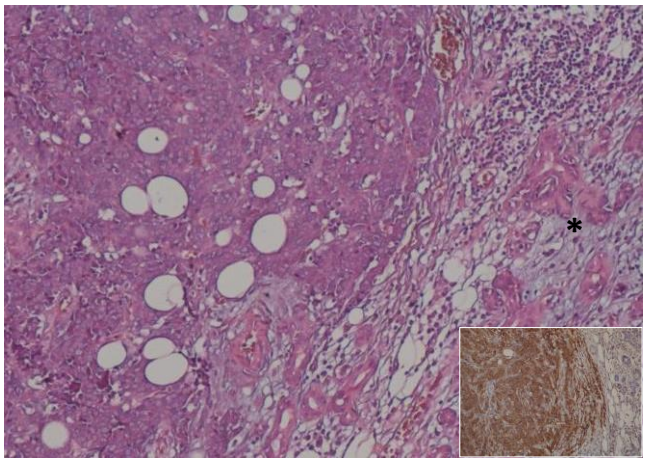

Figure 2 : Pleomorphic cellular infiltrate with small cells scattered in a fibrovascular stroma. Note the parotid gland's acini* (HE x 250). In set: Positive immunohistochemical staining of tumor cells for CD99 (IHC x 100).

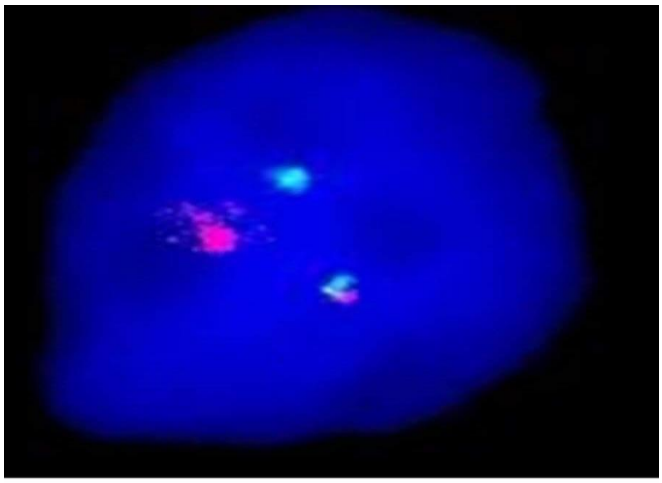

Figure 3: Fluorescence in situ hybridization demonstrated chromosomal rearrangement in the EWSR1 gene region. Cells with t (22q12) revealed one fusion, one orange and one green signal pattern.

Author	Year	Age	Gender	Treatment	Metastases
Jones et al	1995	15	Female	Chemotherapy +radiation therapy	None
Deb et al	1998	60	Female	Surgery	Not available
Deb et al	1998	45	Male	Surgery	None
Helsel et al	2000	38	Female	Surgery	None
Wang et al	2013	2	Male	Surgery+ radiation therapy+ chemotherapy	None
Kalantari et al	2015	26	Male	Chemotherapy+ radiation therapy	None
Present case	2020	33	Female	Surgery+ chemotherapy+ radiation therapy	None

Table 1: Review of reported cases of primary primitive neuroectodermal tumor of the parotid

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Authors contributions:

Yosra Yahyaoui: Guarantor, Concepts,design,literature search, manuscript preparation

Yosr Zenzri : Concepts,design,literature search,manuscript preparation, statistical analysis

Bettaieb Ilhem: definition of intellectual content, Design

Amel Mezlini: Concepts, Definition of intellectual content

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