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CASE REPORT

Mouth metastasis of peripheral primitive neuroectodermal tumor

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We describe a case of oral metastasis of peripheral primitive neuroectodermal tumor (pPNET) in a 68-year-old man, who presented the primary lesion in the chest. Oral metastasis of pPNETs is very rare and we have not found any similar case reported in the English literature. Clinical examination showed an extensive and ulcerated fleshy mass measuring 3.0 × 3.5 cm in the right lower gingivae. Microscopic examination showed sheets of proliferating small, hyperchromatic, round cells. Tumor cells were reactive to neuron-specific enolase (NSE), vimentin and MIC-2 gene by immunohistochemistry, consistent with PNET. The patient died 3 weeks later because of respiratory insufficiency.

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Introduction

Primitive neuroectodermal tumor (PNET) is a term used to describe a category of neoplasms of neuroectodermal origin with variable cell differentiation (Hyun *et al*, 2002). It was first coined for a group of embryonal tumors located in the central nervous system (cPNET). More recently, the PNET concept has been expanded to include histologically similar, peripherally located tumors, referred to as pPNETs or peripheral PNETs (Kao *et al*, 2002).

Peripheral PNETs are rare, affecting mainly children and young adults (Helsel *et al*, 2000). PNETs usually involve bone or soft tissue, the chest and limbs, and only 3% have been reported within the skull and jaws (Jones and McGill, 1995). These tumors are aggressive, with a tendency to recur and to metastasise especially to bone marrow, brain, lungs, and lymph nodes (Helsel *et al*, 2000; Hyun *et al*, 2002).

In the thoracopulmonary region, PNET primarily involves the chest wall, but rarely presents as a primary pulmonary neoplasm (Imamura *et al*, 2000; Kahn *et al*, 2001). The thoracic region is the most common site for recurrent disease, manifested as local chest wall recurrence or pulmonary metastasis. The next most common area involves skeletal, brain, liver, and adrenal glands metastasis (Catalan and Murphy, 1997). In English literature, only eight cases of primary lung PNET have been reported, five of them with follow-up. Of the five patients, only one developed widespread metastasis in the right lung, left adrenal gland and right ovary (Catalan and Murphy, 1997; Imamura *et al*, 2000; Kahn *et al*, 2001).

The PNETs have been increasingly reported in recent years but there are still very few reports of metastatic PNETs, especially to the head and neck region (Chowdhury *et al*, 1990; Helsel *et al*, 2000; Kao *et al*, 2002). We describe a case of oral metastasis of pPNET in an adult patient, who presented the primary lesion in the chest. To the best of our knowledge, this is the first report of an oral metastatic pPNET.

Case report

A 68-year-old man was referred to the Dentistry Section of the Hematology Center, UNICAMP, in August 2002, with complaint of a rapidly growing painful mass on the right lower gingivae lasting 1 month. The patient was diagnosed with chest pPNET 5 months earlier, and was being treated by chemotherapy.

Oral clinical examination revealed an extensive and ulcerated fleshy mass measuring 3.0 × 3.5 cm in the right lower gingivae extending from first bicuspid to the second molar area (Figure 1). Radiographs of the oral cavity showed no bone involvement. With a clinical diagnosis of metastatic pPNET, an incisional biopsy of the mass was taken under local anesthesia. Microscopic examination showed sheets of proliferating small, hyperchromatic, round cells with ovoid nuclei and scarce pink cytoplasm (Figure 2a). Tumor cells were reactive to neuron-specific enolase (NSE), vimentin and

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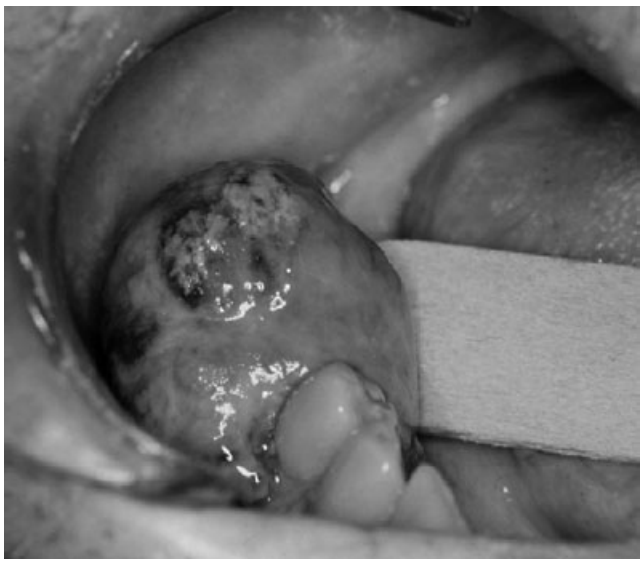

Figure 1 Ulcerated mass on the right mandibular premolar–molar region. The diagnosis was metastatic pPNET of the chest

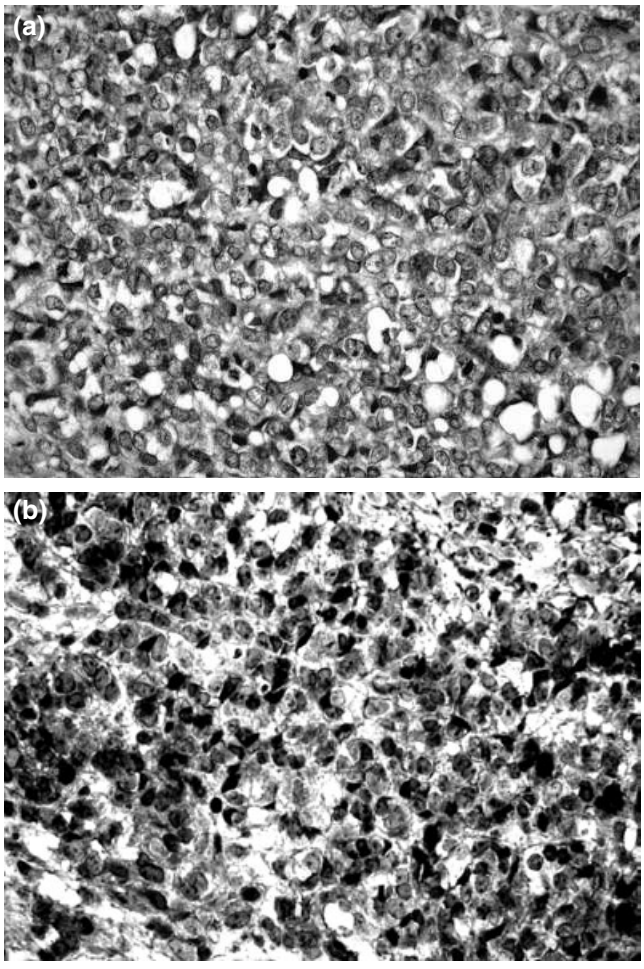

Figure 2 (**a**) Microscopical aspects of mandibular metastasis of pPNET. The tumor is formed by solid sheets or round uniform cells, with scarce pink fibrila to granular cytoplasm (H&E, 100×); (**b**) cells were positive for neuron-specific enolase (NSE), vimentin and MIC-2

MIC-2 gene by immunohistochemistry (Figure 2b), consistent with PNET. The patient died 3 weeks later because of respiratory insufficiency.

Discussion

Primitive neuroectodermal tumor belongs to the specific group of neoplasms that are difficult to distinguish on the basis of cytology and histology alone. Light microscopy of PNETs often reveals small round cells that may form a lobular or pseudorosette pattern, called Homer–Wright rosettes (Helsel *et al*, 2000; Kao *et al*, 2002). Histological differential diagnosis includes rhabdomyosarcoma, Ewing's sarcoma, neuroblastoma, and lymphomas (Jones and McGill, 1995).

There are several antibody neural markers (NSE, S-100, Leu-7, chromogranin, and neurofilaments) which specifically label the PNET cells, and which are negative in Ewing's sarcoma. However, positive immunostaining for vimentin and CD99 (MIC-2 gene) is of particular diagnostic value for differentiating PNET and Ewing's sarcoma from other small round cell tumors (Chowdhury *et al*, 1990; Jones and McGill, 1995; Kao *et al*, 2002).

Peripheral primitive neuroectodermal tumors are most common in children and young adults, however the case reported herein described a 68-year-old patent. There are no previous reports of pPNET metastasis into the mouth; however it should be included in the differential diagnosis of mouth lesions, as illustrated here.

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