Peripheral Primitive Neuroectodermal Tumor- Imaging Appearance
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Citation

Abstract
Peripheral primitive neuroectodermal tumors (PPNET) are rare but highly aggressive tumor of the peripheral non-central nervous system tissue. Thoracopulmonary region is the most common site, however they have been described in other sites as well. We present imaging appearance of three such tumors located in the chest, abdomen and mandible.

INTRODUCTION
Peripheral primitive neuroectodermal tumor (PPNET) is uncommon, and the overall incidence is 1% of all sarcomas. These typically arise outside the central and sympathetic nervous system. The most common locations of PPNETs have been the thoraco-pulmonary region, the retroperitoneal paravertebral soft tissues, the soft tissues of the head and neck, and the intraabdominal and intrapelvic soft tissues and extremities. The diagnosis is based on any hint of neural differentiation at light or electron microscopic level. The rarity of this tumor resulted from diagnosing the most of the PNETs as Ewing’s Sarcoma in the past.

CASE REPORT
CASE No. 1: A 20-year old male patient was admitted to the hospital after a two-month history of cough and a 6 kg weight loss. The patient's initial X-Ray film showed a small pleural effusion. He was treated for pneumonia. Two weeks later a repeat chest X-Ray film revealed a much larger right pleural effusion and the right hemidiaphragm was raised. (Fig 1a) Thoracocentesis revealed an exudative effusion with 100 percent lymphocytes. This effusion recurred in 48 hours. A pleural biopsy was subsequently performed which revealed chronic inflammation. CT scan was performed at this stage which revealed a large, well defined, poorly enhancing mass lesion occupying the left hemithorax inferiorly measuring approximately 12 x 14 cm. There was associated minimal pleural effusion. There was no calcification within the mass. Also no definite areas of necrosis could be demonstrated within the mass lesion. Adjacent bony structures were normal with no evidence of erosion. (Fig 1b) MRI revealed a similarly well-defined mass lesion isointense to muscle on T1-weighted images (Fig 1c) and heterogeneously hyperintense on T2-weighted images (Fig 1d). There were areas suggestive of hemorrhage within the mass lesion. T2-weighted images also revealed areas suggestive of necrosis. There was associated right sided pleural effusion. Mediastinal structures were displaced with no obvious evidence of infiltration. Biopsy revealed small round cell tumor consistent with primitive neuroectodermal tumor.

Figure 1
Figure 1a: Radiograph chest postero-anterior view revealing right sided pleural effusion with raised right hemidiaphragm
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Figure 2
Figure 1b: Sagittal reconstruction of CT images revealing a well-defined heterogeneously enhancing mass lesion involving left hemithorax inferiorly measuring approximately 12 x 14 cm. In addition minimal pleural effusion is seen. There was no calcification within the mass. Also no definite areas of necrosis could be demonstrated within the mass lesion. Adjacent bony structures were normal with no evidence of erosion.

CASE No. 2: An 11 year old female child presented history of painless progressively enlarging mass lesion of approximately 2 months duration. On examination the mass was huge, firm, non tender and fixed originating from mandibular symphysis. The overlying buccal mucosa and neck were normal. CT scan revealed a large 10 x 12 cm mass lesion destroying the body of mandible on left side and invading the roots of the lower teeth. (Fig 2) Incisional biopsy revealed a small round cell tumor consistent with PNET.

Figure 3
Figure 1c: Axial T1-weighted image revealing a well-defined mass lesion isointense to muscle on T1-weighted images. There are hyperintense foci within the mass lesion suggestive of hemorrhage. There was associated right sided pleural effusion. Mediastinal structures are displaced with no obvious evidence of infiltration.

Figure 4
Figure 1d: Axial T2-weighted images of the same patient revealing heterogeneously hyperintense on T2-weighted images with areas suggestive of necrosis.

Figure 5
Figure 2: Reconstructed Panoramic view of CT scan images revealing a large 10 x 12 cm mass lesion destroying the body of mandible on left side and invading the roots of the lower teeth.
CASE No. 3: A 16 year old female patient who presented with vague abdominal pain of 1-2 months duration. On X-ray examination of the abdomen in the supine position there was a large soft tissue density mass lesion overlapping the left iliac bone obscuring the outline of left psoas muscle inferiorly. Ultrasound of the abdomen was performed and it revealed a large hypoechoic left paravertebral mass. On Computed Tomogram (CT) it was seen as a large heterogeneously enhancing hypodense mass measuring 7x 8 cm, in left paravertebral region displacing the psoas anteriorly and partially infiltrating it. (Fig3a) While on Magnetic Resonance Imaging (MRI) it revealed hypointensity on T1-weighted images (Fig 3b) and hyperintensity on T2-weighted images. (Fig 3c). Left psoas was displaced anteriorly with no evidence of infiltration. No evidence of calcification was noted and there was no erosion of adjacent bony structures. FNAC was performed which revealed scattered nests of small tumor cells with round to oval nuclei. The nuclear-cytoplasmic ratio was high, with occasional mitotic figures. There were focal areas of necrosis without rosette formation. The PAS reaction was negative.

**Figure 6**
Figure 3a: Axial CT scan image below the level of aortic bifurcation reveals a large heterogeneously enhancing hypodense mass measuring 7x 8 cm, in left paravertebral region displacing the psoas anteriorly and partially infiltrating it. No evidence of calcification was seen within the lesion. Adjacent vertebra is normal.

**DISCUSSION**
PNETs were first reported by Stout in 1918. Initially they were believed to arise from major nerves. Later reports described these tumours in other anatomic locations as well. Today, the tumor is considered to be a neoplasm of non neural soft tissues that primarily affect the children and adolescents without an apparent gender predilection. These belong to the family of “small round cell blue tumors”
commonly found in the pediatric population. These include neuroblastoma, malignant lymphoma, rhabdomyosarcoma, Ewing's sarcoma, Wilms' tumor and desmoplastic small round cell tumors in addition to PNETs. Central and peripheral primitive neuroectodermal tumors (PNETs) exhibit characteristic immunophenotypical and genetic features that distinguish them from other small round cell tumors. Peripheral PNETs typically express high amounts of the MIC2 antigen (CD99) and exhibit highly characteristic chromosomal translocation between chromosomes 11 and 22. They possess neuronal features with neurosecretory granules on electron microscopy and immunohistochemical characteristics, such as positive staining with neuron specific enolase, which make them a distinct pathologic entity and differentiated them from other round cell tumors such as Ewing's sarcoma and neuroblastoma. 

This tumor can occur at any age, although the peak age incidence is adolescence and young adulthood. There is no sex predilection. In their series of 33 patients Dick et al found the age range to be 0-16 years.

Various reports have described these tumors as large masses with areas of necrosis or hemorrhage, reflecting the aggressive nature of these tumors. Dick et al found that the tumors were typically of soft tissue density on CT with larger tumors (>5 cm) masses tending to be more heterogenous in character. On MRI they were of slightly higher signal than muscles on T1-weighted sequence and all masses were heterogenous on T2W sequences. Calcification was uncommon and generally sparse. These tend to displace adjacent soft tissues structures rather than invade or encase them. These rarely cross the midline. Local or bony invasion at diagnosis was seen. While Kim MS et al found these tumors to be large and aggressive with non-specific imaging appearance. Majority showed heterogenous enhancement with areas of necrosis and hemorrhagic foci. Few of these patients revealed vascular involvement, invasion of adjacent organs and intratumoral cyst and septations. Foci of calcifications were seen in 1 of 10 of their cases. Metastasis was seen to lungs, supraclavicular lymph nodes, mediastinal lymphnodes and pleural fluid in 3 of their series. However, metastasis was seen to lung, pleura, brain, bone, lymphnodes, liver, subcutaneous tissues, kidney and peritoneum in the series reported by Dick et al.

Askin’s tumor is a relatively rare, highly aggressive neoplasm of the thorax which typically occurs in young women, with median survival of eight months. The incidence of peripheral PNET in the abdomen and pelvis, including the retroperitoneum, is about 14% of all peripheral PNETs. Peripheral PNETs of the retroperitoneum have been described in a few publications; it is sporadically reported in the kidney, adrenal gland, and pelvis. Although head and neck region is a rare site of origin for these malignant neoplasms, they have been reported in the skull, orbit, masseter muscle, and maxilla. To the best of our knowledge only 05 other cases of mandibular PNET have been described in the literature so far.

Extraosseous Ewing’s sarcoma (EOES) is an important differential diagnosis of PPNET. The distinction between PPNET and extraosseous Ewing’s sarcoma (EOES) is important because disease free survival is poorer for patients with PPNET than for those with EOES. Both tumors occur most commonly in truncal & paravertebral soft tissues (50% to 60% of cases) and in extremities (25% of cases), although PPNET occurs less commonly in extremities than does EOES, and patients with EOES are generally younger.

CONCLUSION

We present our experience with peripheral PNETs over a period of 05 years. These are rare but highly aggressive tumors which do not have any typical imaging features. Their diagnosis depends on histopathology, immunohistochemistry and electron microscopy. Early recognition is important for proper treatment of these tumors.

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References

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