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FULL PAPER

CT and clinical findings of peripheral primitive neuroectodermal tumour in children

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Objective: To describe the clinical, CT and pathological findings of paediatric peripheral primitive neuroectodermal tumours (pPNETs) to enhance the recognition of these rare tumours.

Methods: The clinical, CT and pathological findings of 18 paediatric patients with pPNETs confirmed by biopsy or surgical pathology were retrospectively reviewed.

Results: The age of these 18 paediatric patients with pPNETs ranged from 4 months to 15 years, with a mean age of 7.7 years. The lesions of these 18 paediatric patients with pPNETs were located in the head and neck ($n = 4$), chest ($n = 2$), abdomen and pelvic cavity ($n = 6$), spine ($n = 3$), ilium ($n = 2$) and femur ($n = 1$). Immunohistochemical examination revealed Homer–Wright rosettes in seven lesions, and 94.4% of lesions showed consistent positive staining for CD99. On plain CT images, the majority of pPNETs showed lesions that were ill-defined (72.2%), irregularly shaped (83.3%), heterogeneous (66.7%) or hypodense masses (94.4%), and together with osteolytic bone destruction when the lesion originated in the bone. Calcifications were found in three lesions. After contrast administration, all soft-tissue

masses were persistently enhanced heterogeneously with various cystic or necrotic regions, and 71.4% of them had linear enhancement. 94.4% of soft-tissue masses showed a moderate degree of enhancement. Seven cases had lymph node metastasis at diagnosis.

Conclusion: Paediatric pPNET can involve any part of the body, and a large, ill-defined, aggressive soft-tissue mass and moderate heterogeneous enhancement with varying cystic regions and linear enhancement, with or without osteolytic bone destruction, on CT images could suggest the diagnosis.

Advances in knowledge: Primitive neuroectodermal tumours constitute a rare type of malignant neuroectodermal tumours that have chromosomal translocations identical to Ewing's sarcoma, and reports about radiological characteristics of this disease in children are insufficient. This study has described the clinical features and CT and pathological findings in 18 paediatric patients diagnosed with pPNETs in different locations, as a way to enhance the recognition of these tumours and help to differentiate from other types of paediatric malignant bone and soft-tissue tumours.

INTRODUCTION

Primitive neuroectodermal tumours (PNETs) are rare, malignant, small, round cell tumours that consist of only 1% of all sarcomas and have highly aggressive biological behaviour and poor prognosis.^{1,2} According to the 2013 WHO classification of tumours of soft tissue and bone, both Ewing's sarcoma (ES) and PNET have a similar gene–phenotype, which can strongly express CD99 and exhibit characteristic chromosomal translocation $t(11;22)(q24;q12)$.³ Hence, they should be viewed as the same tumours, and these tumours have similar clinical appearances, immunohistochemical features, cellular molecular characteristics and prognoses, differing from each other

only in their degree of neural differentiation. Most commonly, PNET occurs in the central nervous system. However, it also sporadically involves extracranial sites. Depending on the location, PNET can be further classified as central and peripheral PNETs (pPNETs); pPNET includes extraosseous ES and Askin's tumour.^{4,5} pPNET can occur anywhere in the body except the central and sympathetic nervous system and the thoracopulmonary region (where it is known as Askin's tumour) which is the most common of these sites.^{6,7} The clinical presentation varies depending on the different sites of involvement, and rapidly growing masses or associated symptoms are the most common findings.

Table 1. Clinical and pathological findings in 18 paediatric patients with peripheral primitive neuroectodermal tumour

Case	Age	Sex	Location	Size (cm)	CD99	Vimentin	NSE	S-100 protein	Syn	CK	Myogenin
1	4 months	Female	Carotid sheath	4.8	+	+	+	-	+	-	-
2	6 years	Male	Parotid gland	6.4	+	+	-	-	-	-	-
3	1.5 years	Female	Maxillae	4.4	+	+	+	-	+	-	-
4	14 years	Female	Submaxilla	4.2	+	-	+	-	-	-	-
5	15 years	Female	Pleura	15.4	+	-	-	-	+	-	-
6	12 years	Male	Pleura	9.8	+	-	-	-	+	-	-
7	3 years	Male	Adrenal gland	6.3	+	+	-	-	-	-	-
8	7 years	Female	Pelvic cavity	8.0	+	+	-	-	-	-	-
9	5 years	Male	Pancreas	6.4	+	-	+	-	-	-	-
10	4 years	Male	Abdomen and pelvic cavity	13.3	+	+	-	-	-	-	-
11	5 years	Male	Pancreas	7.9	+	-	-	-	+	-	-
12	1 year	Female	Kidney	10.1	+	+	+	-	-	-	-
13	3 years	Female	Sacral vertebrae	2.2	+	+	+	-	-	-	-
14	10 years	Male	Sacral vertebrae	4.0	+	-	-	+	-	-	-
15	10 years	Female	Lumbar vertebrae	3.2	+	+	-	-	-	-	-
16	15 years	Male	Femur	9.5	+	-	+	-	-	-	-
17	13 years	Female	Ilium	6.4	-	+	+	-	-	-	-
18	14 years	Female	Ilium	5.7	+	+	-	-	-	-	-

+, positive result; -, negative result; CK, cytokeratin; NSE, neuron-specific enolase; Syn, synaptophysin.

Figure 1. Primitive neuroectodermal tumour of the left carotid sheath in a 4-month-old female. On plain CT image (a), the lesion exhibits a round hypodense mass with small lamellar lower density; on contrast-enhanced CT images (b), the lesion was persistent heterogeneously enhanced with an area of cyst (arrowheads), and multiple enlarged cervical lymph nodes were confirmed as metastasis (arrowheads).

Although pPNET can affect persons of any age group, the peak incidence is in children and young adults; and it is usually located in the thorax, head and neck, and abdomen.^{8–10} However, the diagnosis of pPNET is still a challenge to the paediatrician because of the lack of typical presentation and low incidence. The clinical and pathological characteristics of pPNET are definite, but the radiological features have been reported in only a few studies, and in these few studies, most of them are cases arising in the common locations and are reported in the form of case reports.^{6,9–16} This report describes the clinical features, and CT and pathological findings in 18 paediatric patients diagnosed with pPNETs in different locations, as a way to enhance the recognition of these tumours and help differentiate these from the other types of paediatric malignant bone and soft-tissue tumours.

METHODS AND MATERIALS

Patient population

The clinical and imaging records of 18 paediatric patients confirmed as pPNET between January 2011 and August 2013 were retrospectively reviewed. All patients had pre-operative CT

records. There were 10 females and 8 males, with an age range from 4 month to 15 years (mean: 7.7 years old). Of these 18 paediatric patients with pPNETs, 10 presented with persistent pain of the involved sites; 5 patients had a large mass; and there were 3 patients with some rare manifestations, such as icteric sclera, exophthalmos and difficulties of defaecation. The duration of the symptoms was between several days and 12 months. There were 18 patients in our study, 9 cases were confirmed by surgical pathology, 9 cases were confirmed by core needle pathology, and core biopsy was performed with 18-gauge puncture needles under CT guidance. All the pathologic specimens were reviewed by pathologists for routine haematoxylin and eosin staining and immunohistochemical staining for immunological markers. Immunological markers that were assessed included CD99, neuron-specific enolase, vimentin, S-100 protein, cytokeratin, synaptophysin, leukocyte common antigen, epithelial membrane antigen and myogenin. The diagnosis of pPNET was made when there was evidence showing that cells were small, round, arranged with or without rosettes, with neural differentiation, as evidenced by positive staining for two or more types of neuron-specific antibodies.

Figure 2. Primitive neuroectodermal tumour of the right parotid gland in a 6-year-old male. On plain CT image (a), the lesion shows an irregular hypodense mass with small lamellar lower density; on enhanced CT image (b), the lesion was enhanced heterogeneously with an area of cystic and linear enhancement.

Figure 3. Primitive neuroectodermal tumour of the right maxillae in an 18-month-old female. Axial and coronal plain CT image shows osteolytic destruction of the right maxilla and ill-defined margin, irregular hypodense mass (a), and the lesion involves the right accessory nasal sinuses and orbital cavity (b; arrowheads).

Imaging techniques

All 18 paediatric patients underwent CT scans using a SOMATOM® Sensation Cardiac 64-slice CT (Siemens Medical Systems, Erlangen, Germany) helical scanner. The scanning parameters were as follows: 5-mm slice thickness reconstructions, 45-cm field of view, 120 kV voltage, 200 mA and 512×512 matrix. Three-dimensional CT images were reconstructed using InSpace software (the Siemens LEONARDO Workstation). 4 patients underwent plain CT scanning, and the other 14 patients underwent plain and contrast-enhanced CT scanning. A non-ionic iodinated contrast agent, Iopromide (Ultravist®, Bayer Schering Pharma AG, Berlin, Germany), was administered to the patients who had enhanced CT scanning at a rate of 3 ml s^{-1} , followed by a 20-ml saline flush, and the dose of contrast agent was calculated according to the weight of the patients (1.2 ml kg^{-1}). Arterial phase and venous phase images were obtained; imaging was performed 60 and 105 s after contrast material injection.

Imaging interpretation

All the patients' images were reviewed independently by two experienced radiologists with 5 or more years' of radiological experience. The radiologists were blinded to the final pathological results, and the final assessment was reached by consensus through discussion. The aspects of the tumour that were described on CT included tumour location, size, morphology, margin (well-defined or ill-defined), CT density (hypo-, iso- or hyperdense in relation to the adjacent muscle density), calcification, contrast enhancement appearances (homogeneous or heterogeneous), lesion texture (homogeneous or heterogeneous

or necrosis), and the conditions of the adjacent tissue structures and distant metastasis at diagnosis. We then compared the image findings with the pathological results.

RESULTS

The clinical and pathological findings of the 18 paediatric patients with pPNET are shown in Table 1. The tumours were located in the head and neck ($n = 4$), chest ($n = 2$), abdomen and pelvis ($n = 6$), spine ($n = 3$), ilium ($n = 2$) and femur ($n = 1$). Of the four cases with lesion sites in the head and neck, the locations included the carotid sheath (Figure 1), parotid gland (Figure 2), submaxilla and maxillae (Figure 3), and the size of them ranged from 4.2 to 6.4 cm (mean: 5.0 cm). Two pPNET lesions occurred at the pleura in our study (Figure 4), with a maximal diameter of 9.8 and 15.4 cm (mean: 12.6 cm), respectively. In total, six cases had the locations in the abdomen and pelvis, including two abdomen and pelvis without definite origin and two pancreas (Figures 5 and 6), one adrenal gland and one kidney (Figures 7 and 8); and the size ranged from 6.3 to 13.3 cm (mean: 8.7 cm). There were three lesions arising in the spine with a size range from 2.2 to 4.0 cm (mean: 3.1 cm), including two sacral vertebrae and one lumbar vertebra (Figure 9). The two lesions arising in the ilium and one occurring in the femur (Figure 10) had the size range from 5.7 to 9.5 cm (mean: 7.2 cm). Histologically, all the tumours predominately consisted of small, round, undifferentiated cells, and Homer–Wright rosettes were observed in seven lesions; and the immunohistochemical examination revealed 94.4% (17/18) of lesions showing consistent positive staining for CD99. The

Figure 4. Primitive neuroectodermal tumour of the right pleura in a 12-year-old male. Contrast-enhanced CT demonstrates a huge moderately enhanced cystic-solid mass with multiple linear enhancements (a, b).

Figure 5. Primitive neuroectodermal tumour of the abdomen and pelvis in a 4-year-old male. Plain CT image (a) shows a huge hypodense mass; and contrast-enhanced CT demonstrates a marked enhanced cystic-solid mass with multiple areas of cyst (b; white arrowheads) and linear enhancements (b; black arrowheads).

percentage of other markers, specifically vimentin, neuron-specific enolase, synaptophysin and S-100 protein, were 61.1% (11/18), 44.4% (8/18), 27.8% (5/18) and 5.6% (1/18), respectively. Cytokeratin and myogenin were both negative in all cases.

All 18 cases underwent CT scanning, including 4 plain CT scanning and 14 plain and contrast-enhanced scanning; the CT findings are listed in Table 2. Of these 18 pPNET lesions, there were 8 lesions arising in a bony location and 10 arising in extraosseous tissue. In spite of the locations of the lesions, soft-tissue mass was found in all pPNET lesions on plain CT images; and the majority of them showed lesions that were ill-defined (72.2%, 13/18), irregularly shaped (83.3%, 15/18), heterogeneous (66.7%, 12/18) or hypodense mass (94.4%, 17/18). For the eight lesions arising in the bone, osteolytic bone destruction was also found, and the normal bone structure was replaced by an irregular hypodense mass (Figures 3, 9 and 10). Of the 18 pPNET lesions, 12 lesions showed heterogeneously dense masses with multiple lamellar lower densities (Figures 1a, 2a, 6a and 8a). There were three well-defined round ($n = 1$) or oval ($n = 2$) hypodense masses in our study, and one case occurred at the carotid sheath and two at the abdomen and pelvis (Figures 1a, 6a). Calcification was found in three lesions of these 18 patients (16.7%, 3/18), showing linear or punctiform shapes (Figure 6a), and the lesions were located at the pleura, pancreas and adrenal gland. After contrast agent

administration, all soft-tissue masses were persistently enhanced heterogeneously, with varying cystic or necrotic regions (Figures 1, 2, 4–10), and 71.4% (10/14) of these linear enhancements could be found (Figures 1, 2, 4–8). Small arteries that originated from the abdominal aorta could be found in all four pPNET lesions arising in the organs (including two pancreas, one adrenal gland and one kidney) in our study (Figures 6d, 7c and 8b). Of 14 enhanced mass lesions, 9 lesions had ill-defined margins and 5 lesions had well-defined margins, which had no changes compared with the margins of lesions displayed on the plain CT images. Except for one lesion occurring at abdomen and pelvis exhibiting marked enhancement (Figure 5b), all soft-tissue masses showed a moderate degree of enhancement.

Of these 18 patients with pPNET, 7 cases had lymph node metastasis at diagnosis, including 3 cervical lymph nodes, 3 retroperitoneal lymph nodes and 1 axillary node (Figures 1 and 7d). Three lesions occurring in the spine showed an irregular mass extending to the adjacent spinal canal (Figure 9); and one lesion arising in the maxillae had an invasion of accessory nasal sinuses and orbital cavity (Figure 3). Tumour thrombus extending into the renal vein and inferior vena cava was noted in the renal-region pPNET. Besides, pleural fluid and ascites were found in four patients; focal pulmonary atelectasis and intra-hepatic duct dilatation were complicated with two pleural lesions and a pancreatic lesion, respectively.

Figure 6. Primitive neuroectodermal tumour of the pancreas in a 5-year-old male. The plain CT image (a) shows multiple linear, punctiform calcifications in an irregular hypodense mass (arrowheads); contrast-enhanced images (b–d) exhibit an enhanced cystic-solid mass with multiple areas of cyst and linear enhancements. Small artery that originated from the abdominal aorta could be found among the lesions (d; arrowhead).

Figure 7. Primitive neuroectodermal tumour of the right adrenal gland in a 3-year-old male. Plain CT image (a) shows an irregular heterogeneous hypodense mass in the right adrenal gland region; axial and coronal enhanced CT images (b, c) demonstrate moderate enhanced cystic-solid mass, and small arteries that originated from the abdominal aorta could be found (c; arrowheads) among the lesions. Multiple enlarged retroperitoneal lymph nodes are confirmed as metastasis (d).

DISCUSSION

The concept of PNET was raised firstly by Stout in 1918 by reporting a 42-year-old male with ulnar nerve tumours,¹⁷ and in 1921, Ewing¹⁸ discussed a 14-year-old male with osteoclasia in the ulna and suggested the name Ewing sarcoma. Its actual existence was debated until Hart and Earle¹⁹ described a group of unspecialized small-cell tumours with the name of PNET. In fact, these two tumours were derived from the same origin of primitive neuroectoderm and have the same chromosomal translocations. The current opinion is that as both ES and PNETs have a similar phenotype, they should be viewed as the same tumour, differing from each other only in their degree of neural differentiation. Clinically, pPNET mainly appear as rapidly growing masses, and the symptoms, such as pain and compression, are usually due to the mass. pPNETs occur at any age but mainly affect children and young adults.^{6,9-16,20} In our series, the age range was from 4 months to 15 years (mean age: 7.7 years), which was consistent with the literature,^{6,10,20} although there were infant patients reported in other studies.^{8,9} pPNET can occur at any location, of which the thoracopulmonary region, head and neck, and extremities are the most common sites.^{9,10,14} In our series, the sites of tumours were in the abdomen and pelvis ($n = 6$), head and neck ($n = 4$), spine ($n = 3$), ilium ($n = 2$), femur ($n = 1$) and chest ($n = 2$); which were a little different from the reports above.^{9,10,14} In our study, the rare locations of carotid sheath, parotid gland, pancreas, adrenal gland and kidney were described. Comparing with the size of pPNET in different locations, we found that the size of the lesions arising in the spine (<5 cm) was smaller than that of

the lesions arising in organs, pleura, soft-tissue space and extremities; and tumours arising from the chest or abdominopelvic cavity were larger than those arising elsewhere, which was consistent with that of a previous report.⁹ This may be due to the rapid growth and availability of more growth space for the locations mentioned above. Generally, the prognosis of pPNET is unfavourable.^{8,9} A combination of adequate surgical resection, effective multiagent chemotherapy and concurrent radiotherapy, if possible, is necessary to improve survival. Unfortunately, we did not follow up further after surgery in our study. Histologically, pPNETs predominately consist of undifferentiated, small, round cells, and Homer–Wright rosettes were observed in seven lesions in our study. The differential diagnoses of this tumour include malignant lymphoma, small-cell (neuroendocrine) carcinoma, rhabdomyosarcoma and other types of paediatric malignant bone and soft-tissue tumours. For differential diagnosis, immunohistochemical analyses should be performed. In our series, the immunohistochemical examination revealed 94.4% (17/18) lesions stained strongly for CD99. This is consistent with other reports in the literature.^{3,21}

Diverse radiological findings have been reported in patients with pPNETs;^{6,7,9-16,22} but for paediatric pPNETs, most of them are cases arising in common locations and are reported in the form of case reports.^{6,9-16} In our series, the CT findings of 18 paediatric pPNETs were described, and the case number was less than that of the study of Dick et al,⁹ which reported 25 young patients with pPNET having CT scans. In our study, both common and uncommon locations of pPNETs were included.

Figure 8. Primitive neuroectodermal tumour of the left kidney in a 1-year-old female. Plain CT image (a) shows a huge ill-defined, heterogeneous hypodense mass with multiple lamellar lower density in the left renal region (a); on enhanced CT images (b, c), the lesion was enhanced persistently heterogeneously with multiple areas of cyst (c; arrowheads) and linear enhancement, and small arteries that originated from the abdominal aorta could be found (b; arrowheads); tumour thrombus extending into inferior vena cava (d; arrowheads).

Figure 9. Primitive neuroectodermal tumour of the sacral vertebra in a 10-year-old male. Axial and sagittal enhanced CT image exhibits osteolytic destruction of the bone and an ill-defined margin, irregular heterogeneously enhanced soft mass extending to the adjacent spinal canal (a, b; arrowheads).

Of our 18 pPNET lesions, there were 8 lesions arising in a bony location and 10 originating in extraosseous tissue. In spite of the locations of the lesions, soft-tissue mass was found in all pPNET lesions on CT images, which was consistent with that of previous reports.^{6,7,9–16,22} In our study, the majority of pPNET lesions showed as ill-defined, irregularly shaped, heterogeneous, hypodense mass on plain CT image accompanied with or without osteolytic bone destruction and persistent moderate heterogeneous enhancement with varying cystic or necrotic regions and linear enhancements after the contrast agent administration. The cystic or necrotic changes on CT images may be due to the rapid growth and lack of blood supply in these tumours. Generally speaking, the areas of cysts or necrosis were more commonly seen in larger tumours, and the appearances of linear enhancements on contrast-enhanced images may have some relationship with the feeding vessels. Small feeding arteries that originated from the abdominal aorta could be found in all four pPNET lesions arising in the organs (including two lesions arising in the pancreas, and one arising in the kidney and one adrenal gland pancreas) in our study, and a similar report was not found in the literature.^{6,7,9–16,22} This may explain why these tumours grow rapidly and are easy to show cystic or necrotic changes; besides, these feeding vessels could help locate the origin of the lesions. Although calcifications are rare in pPNET,^{9,10,22,23} calcifications were found in three lesions in our study showing linear or punctiform shapes, and the lesions were located in the site of the pleura, pancreas and adrenal gland. Hence, in regard to a huge soft-tissue mass with calcifications in children or adolescents, the diagnosis of pPNET should be considered, except when there is a diagnosis of childhood

blastoma, such as neuroblastoma, nephroblastoma, pancreatoblastoma, hepatoblastoma or teratoma. For lesions originating in the bone, periosteal reaction, osteosclerosis and calcifications were not found in our series. These are useful for the differential diagnosis from malignant primary bone tumours, such as osteosarcomas and chondrosarcomas,²⁴ but the differential diagnosis becomes more difficult when lesions of bone metastasis with the features of a soft-tissue mass and osteolytic bone destruction are involved if no primary malignant tumour has been found. Hence, a differential diagnosis should be made between this rare tumour and other types of paediatric malignant bone and soft-tissue tumours.

The lesions of pPNET are commonly aggressive and readily spread to the neighbouring soft tissues and distant organs. In our 18 cases of paediatric pPNETs, 7 cases had lymph node metastasis at diagnosis; 3 lesions occurring at the spine showed an irregular mass extending to the adjacent spinal canal and 1 lesion arising in the maxillae had accessory nasal sinuses and orbital cavity involved. These findings were consistent with that of previous reports;^{9–11,13} however, lymph node metastasis was more frequently found in our study than that of the above reports. Tumour thrombus extending into the renal vein and inferior vena cava was found in a renal-region pPNET in our study, which was consistent with the report of Chu et al.¹³ Common sites of metastases in pPNETs are the bones and lungs. However, in our series, distant metastases cannot be commented upon because clinical follow-up was not conducted. Hence, further studies should focus on survival analysis and should include a larger study population.

Figure 10. Primitive neuroectodermal tumour of the left ilium in a 15-year-old female. Plain CT image shows osteolytic destruction of the bone and an ill-defined margin, irregular hypodense mass (a). Enhanced CT image (b) demonstrates the lesion heterogeneously enhanced with multiple areas of cystic changes.

Table 2. CT Findings in 18 paediatric patients with peripheral primitive neuroectodermal tumour

Case	Scanning mode	Shape	Density	Margin	Homogeneity	Calcification	Enhanced features			Metastasis/ invasion
							Margin	Homogeneity	Degree	
1	Plain and enhanced	Round	Hypo	Well-defined	Homo	No	Well-defined	Hetero; cystic; linear enhancement	Persistent; moderate	Cervical lymph nodes
2	Plain and enhanced	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	No	Ill-defined	Hetero; cystic; linear enhancement	Persistent; moderate	Cervical lymph nodes
3	Plain	Irregular	Hypo	Ill-defined	Homo	No				Accessory nasal sinuses; orbital cavity
4	Plain	Irregular	Hypo	Ill-defined	Homo	No				Cervical lymph nodes
5	Plain	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	Punctiform				No
6	Plain and enhanced	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	No	Ill-defined	Hetero; cystic; linear enhancement	Persistent; moderate	Axillary lymph nodes
7	Plain and enhanced	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	Punctiform	Ill-defined	Hetero; cystic; linear enhancement	Persistent; moderate	Retroperitoneal lymph nodes
8	Plain and enhanced	Oval	Iso	Well-defined	Hetero; cystic-solid	No	Well-defined	Hetero; cystic; linear enhancement	Persistent; moderate	No
9	Plain and enhanced	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	No	Ill-defined	Hetero; cystic	Persistent; moderate	Retroperitoneal lymph nodes
10	Plain and enhanced	Oval	Hypo	Well-defined	Hetero; cystic-solid	No	Well-defined	Hetero; cystic; linear enhancement	Marked	No
11	Plain and enhanced	Irregular	Hypo	Well-defined	Hetero; cystic-solid	Multiple, linear and punctiform	Well-defined	Hetero; cystic; linear enhancement	Persistent; moderate	No
12	Plain and enhanced	Irregular	Hypo	Well-defined	Hetero ; cystic-solid	No	Well-defined	Hetero; cystic; linear enhancement	Persistent; moderate	Retroperitoneal lymph nodes

(Continued)

Table 2. (Continued)

Case	Scanning mode	Shape	Density	Margin	Homogeneity	Calcification	Enhanced features			Metastasis/ invasion
							Margin	Homogeneity	Degree	
13	Plain and enhanced	Irregular	Hypo	Ill-defined	Homo	No	Ill-defined	Hetero; cystic	Persistent; moderate	Sacral vertebrae destruction instead by soft tissue mass
14	Plain and enhanced	Irregular	Hypo	Ill-defined	Homo	No	Ill-defined	Hetero; cystic	Persistent; moderate	Sacral vertebrae destruction instead by soft-tissue mass
15	Plain	Irregular	Hypo	Ill-defined	Homo	No				Lumbar vertebrae destruction instead by soft-tissue mass
16	Plain and enhanced	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	No	Ill-defined	Hetero; cystic; linear enhancement	Persistent; moderate	Femur destruction instead by soft-tissue mass
17	Plain and enhanced	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	No	Ill-defined	Hetero; cystic	Persistent; moderate	Ilium destruction instead by soft-tissue mass
18	Plain and enhanced	Irregular	Hypo	Ill-defined	Hetero; cystic-solid	No	Ill-defined	Hetero; cystic; linear enhancement	Persistent; moderate	Ilium destruction instead by soft-tissue mass

hetero, heterogeneity; homo, homogeneity; hypo, hypodense; iso, isodense.

In summary, paediatric pPNET can involve any part of the body. The diagnosis of pPNET should be considered when CT images show a large, ill-defined, aggressive cystic-solid

mass with persistent moderate enhancement, with or without osteolytic bone destruction, in children and adolescent patients.

REFERENCES

1. Tsokos M, Alaggio RD, Dehner LP, Dickman PS. Ewing sarcoma/peripheral primitive neuroectodermal tumor and related tumors. *Pediatr Dev Pathol* 2012; **15** (Suppl. 1): 108–26. doi: <http://dx.doi.org/10.2350/11-08-1078-PB.1>
2. Sirivella S, Gielchinsky I. Treatment outcomes in 23 thoracic primitive neuroectodermal tumours: a retrospective study. *Interact Cardiovasc Thorac Surg* 2013; **17**: 273–9. doi: <http://dx.doi.org/10.1093/icvts/ivt161>
3. Carvajal R, Meyers P. Ewing's sarcoma and primitive neuroectodermal family of tumors. *Hematol Oncol Clin North Am* 2005; **19**: 501–25. doi: <http://dx.doi.org/10.1016/j.hoc.2005.03.004>
4. Dehner LP. Peripheral and central primitive neuroectodermal tumors. A nosologic concept seeking a consensus. *Arch Pathol Lab Med* 1986; **110**: 997–1005.
5. Kleihues P, Louis DN, Scheithauer BW, Rorke LB, Reifenberger G, Burger PC, et al. The WHO classification of tumors of the nervous system. *J Neuropathol Exp Neurol* 2002; **61**: 215–25. doi: <http://dx.doi.org/10.1093/jnen/61.3.215>
6. Khong PL, Chan GC, Shek TW, Tam PK, Chan FL. Imaging of peripheral PNET: common and uncommon locations. *Clin Radiol* 2002; **57**: 272–7. doi: <http://dx.doi.org/10.1053/crad.2001.0807>
7. Hari S, Jain TP, Thulkar S, Bakhshi S. Imaging features of peripheral primitive neuroectodermal tumours. *Br J Radiol* 2008; **81**: 975–83. doi: <http://dx.doi.org/10.1259/bjr/30073320>
8. Kimber C, Michalski A, Spitz L, Pierro A. Primitive neuroectodermal tumours: anatomic location, extent of surgery, and outcome. *J Pediatr Surg* 1998; **33**: 39–41. doi: [http://dx.doi.org/10.1016/S0022-3468\(98\)90357-8](http://dx.doi.org/10.1016/S0022-3468(98)90357-8)
9. Dick EA, McHugh K, Kimber C, Michalski A. Imaging of non-central nervous system primitive neuroectodermal tumours: diagnostic features and correlation with outcome. *Clin Radiol* 2001; **56**: 206–15. doi: <http://dx.doi.org/10.1053/crad.2000.0614>
10. Zhang WD, Chen YF, Li CX, Zhang L, Xu ZB, Zhang FJ. Computed tomography and magnetic resonance imaging findings of peripheral primitive neuroectodermal tumors of the head and neck. *Eur J Radiol* 2011; **80**: 607–11. doi: <http://dx.doi.org/10.1016/j.ejrad.2011.02.008>
11. Schulman H, Newman-Heinman N, Kurtzbar E, Maor E, Zirkun H, Laufer L. Thoracoabdominal peripheral primitive neuroectodermal tumors in childhood: radiological features. *Eur Radiol* 2000; **10**: 1649–52. doi: <http://dx.doi.org/10.1007/s003300000367>
12. Sunil I, Bond SJ, Nagaraj HS. Primitive neuroectodermal tumor of the sternum in a child: resection and reconstruction. *J Pediatr Surg* 2006; **41**: e5–8. doi: <http://dx.doi.org/10.1016/j.jpedsurg.2006.07.013>
13. Chu WC, Reznikov B, Lee EY, Grant RM, Cheng FW, Babyn P. Primitive neuroectodermal tumour (PNET) of the kidney: a rare renal tumour in adolescents with seemingly characteristic radiological features. *Pediatr Radiol* 2008; **38**: 1089–94. doi: <http://dx.doi.org/10.1007/s00247-008-0971-1>
14. Yeh CH, Yeow KM, Chu SY, Pan KT, Hung CF, Hsueh S, et al. *Imaging Findings Mandibular Primitive Neuroectodermal Tumour: A Report a Rare Case Review Literature*. 2011; **40**: 451–6. doi: <http://dx.doi.org/10.1259/dmfr/92169329>
15. Song HC, Sun N, Zhang WP, Huang CR. Primary Ewing's sarcoma/primitive neuroectodermal tumor of the urogenital tract in children. *Chin Med J* 2012; **125**: 932–6.
16. Mao Y, Sang X, Liang N, Yang H, Lu X, Yang Z, et al. Peripheral primitive neuroectodermal tumors arising in the pancreas: the first case report in Asia and a review of the 14 total reported cases in the world. *Hepatobiliary Surg Nutr* 2013; **2**: 51–60. doi: <http://dx.doi.org/10.3978/j.issn.2304-3881.2012.08.01>
17. Stout AP. A tumor of the ulnar nerve. *Proc N Y Soc Pathol* 1918; **12**: 2–12.
18. Ewing J. Diffuse endothelioma of bone. *Proc N Y Soc Pathol* 1921; **21**: 17–24.
19. Hart MN, Earle KM. Primitive neuroectodermal tumors of the brain in children. *Cancer* 1973; **32**: 890–7. doi: [http://dx.doi.org/10.1002/1097-0142\(197310\)32:4<890::AID-CNCR2820320421>3.0.CO;2-O](http://dx.doi.org/10.1002/1097-0142(197310)32:4<890::AID-CNCR2820320421>3.0.CO;2-O)
20. Ibarburen C, Haberman JJ, Zerhouni EA. Peripheral primitive neuroectodermal tumors. CT and MRI evaluation. *Eur J Radiol* 1996; **21**: 225–32. doi: [http://dx.doi.org/10.1016/0720-048X\(95\)00731-5](http://dx.doi.org/10.1016/0720-048X(95)00731-5)
21. de Alava E, Gerald WL. Molecular biology of the Ewing's sarcoma/primitive neuroectodermal tumor family. *J Clin Oncol* 2000; **18**: 204–13.
22. Gupta P, Hari S, Thulkar S. Imaging spectrum of peripheral primitive neuroendocrine tumours. *Singapore Med J* 2013; **54**: 463–2. doi: <http://dx.doi.org/10.11622/smedj.2013155>
23. Li X, Zhang W, Song T, Sun C, Shen Y. Primitive neuroectodermal tumor arising in the abdominopelvic region: CT features and pathology characteristics. *Abdom Imaging* 2011; **36**: 590–5. doi: <http://dx.doi.org/10.1007/s00261-010-9655-z>
24. Masciocchi C, Sparvoli L, Barile A. Diagnostic imaging of malignant cartilage tumors. *Eur J Radiol* 1998; **27** (Suppl. 1): S86–90. doi: [http://dx.doi.org/10.1016/S0720-048X\(98\)00048-5](http://dx.doi.org/10.1016/S0720-048X(98)00048-5)