

Paraspinal primitive neuroectodermal tumour (PNET) of the chest

C Minné, MB ChB, Dip EC (SA)
N Khan, MB BS, FCRad (D) SA

Department of Diagnostic Radiology, University of Limpopo, Medunsa Campus

Primitive neuroectodermal tumours (PNETs) are tumours from pluripotent neural crest cells¹⁻⁵ and may occur in the central or peripheral nervous system.⁴ PNET must be distinguished from other small round cell tumours like Ewing's sarcoma (ES), extrasosseous Ewing's sarcoma, neuroblastoma, lymphoma, rhabdomyosarcoma and small-cell carcinoma.^{3,4,6,7} Although it can occur at any age, PNET occurs more frequently in adolescents and young adults,^{2,3,6} with a median age of 17 years.^{3,6}

Case report

A 20-year-old black woman presented with a 3-month history of lower back pain, a 2-week history of paraplegia and paraparesis, and 1-week history of loss of sphincter control. A supine chest radiograph (CXR) showed a large mass in the right hemithorax (Fig. 1); this posterior mass could be confirmed on the lateral lumbar spine X-ray. The T11 interpedicular distance was increased and the pedicles were flattened on the anteroposterior (AP) lumbar spine X-ray. Computed tomography (CT) (Fig. 2) and magnetic resonance imaging (MRI) (Figs 3 - 5) showed a well-defined lobulated paraspinal mass in the right hemithorax measuring 18 x 11 x 16 cm. The mass involved most of the lower chest cavity and extended into the spinal canal displacing and compressing the spinal cord anteriorly and to the left (Figs 3 and 4) and expanding

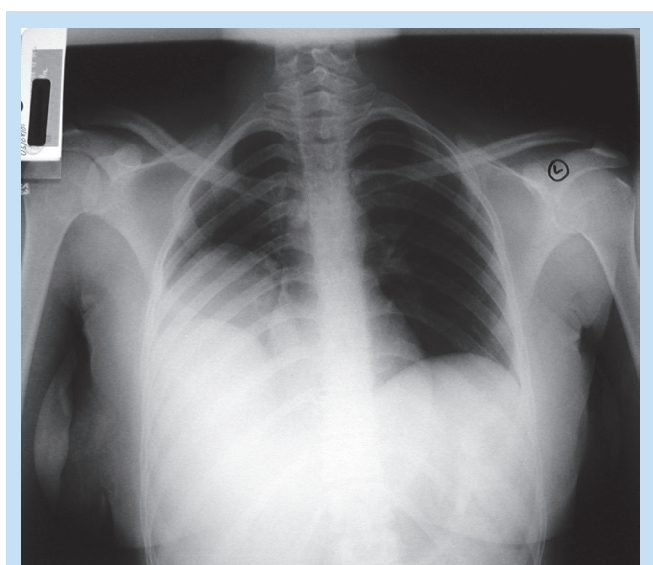


Fig. 1. A supine CXR shows a large mass in the right hemithorax.

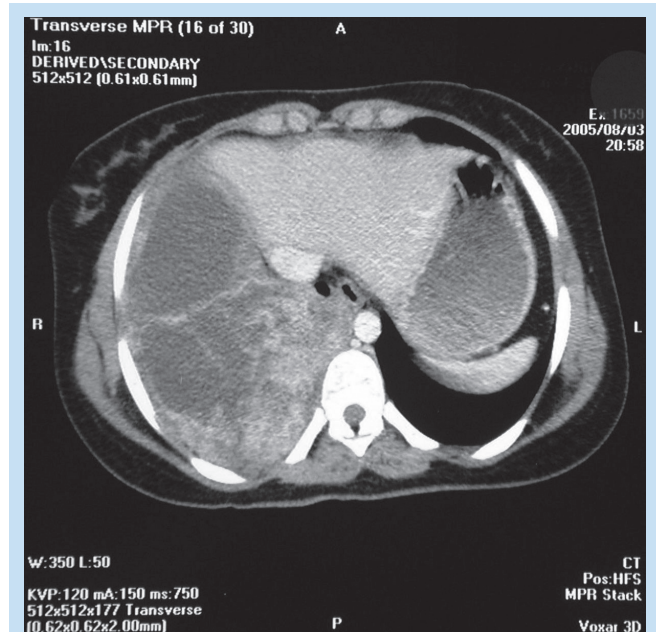


Fig. 2. The post-contrast CT image through the lower chest and upper abdomen demonstrates heterogeneous enhancement of the mass.

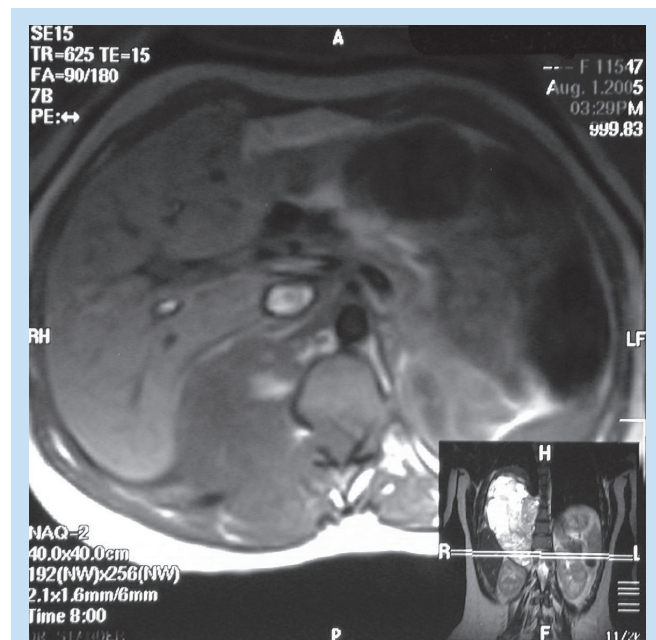


Fig. 3. This T1-weighted axial image shows a right paraspinal mass isointense to muscle. Haemorrhage can be seen as a hyperintense area in the mass. Invasion of the chest wall and erector spinae muscles is seen on the right. The mass extends into the spinal canal via the intervertebral foramen with subsequent widening of the foramen. The spinal cord is compressed and displaced anteriorly and to the left.

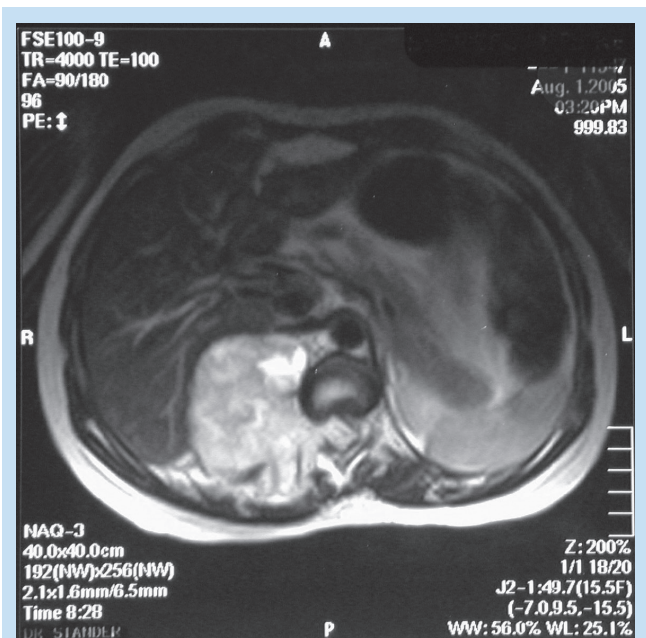


Fig. 4. A T2-weighted image on the same level as Fig. 3 shows a hyperintense heterogeneous mass. The invasion of the chest wall and spinal canal with widened intervertebral foramen and displaced spinal cord is clearly seen.

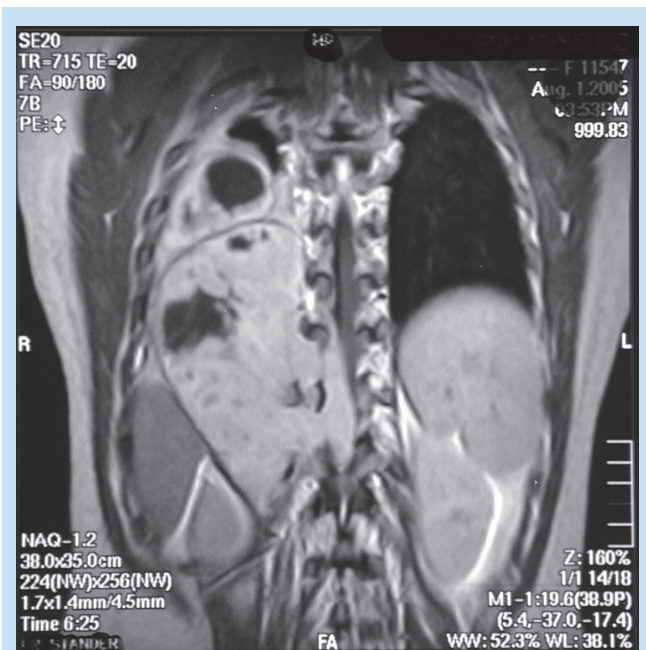


Fig. 5. A coronal post-contrast T1-weighted sequence. The mass enters the spinal canal via numerous neural foramina.

the intervertebral foramina (Figs 3 and 4). The superior portion of the mass had a large cystic component (Fig. 5). The diaphragm could be seen clearly and was displaced antero-inferiorly by the mass, which was seen superiorly to it (Fig. 5). No infiltration of the diaphragm could be demonstrated. The right kidney was subsequently also displaced antero-inferiorly. Infiltration of the chest wall and erector spinae muscles was

well demonstrated on both CT and MRI (Figs 3 and 4). The right ribs were splayed and intervertebral foraminae widened but no bony erosion was seen. No calcification was demonstrated on either CT or MRI, but haemorrhage was seen on MRI (Figs 3 and 4). Regional lymphadenopathy was not seen. The lesion was hypodense to muscle (26 HU) on CT and showed inhomogeneous enhancement post contrast (Fig. 2). On T1-weighted images the mass was isointense to muscle (Fig. 3) and enhancement post gadolinium was heterogeneous (Fig. 5). T2-weighted images showed a hyperintense mass and the cystic component was well demonstrated (Fig. 4). No lung or bony metastases were seen on CT and Tc 99m-MDP bone scan respectively. At surgery an extradural mass was found. The spinal canal was decompressed and a laminectomy was performed on T10-L1, and pedicular screws were inserted to stabilise this region. Histological examination revealed a tumour of small round cells with hyperchromatic nuclei and pale to clear cytoplasm. A few Flexner-Wintersteiner rosettes were seen. Immunohistochemistry revealed the following: S100 was diffuse strong positive, vimentin was positive, CD99 had focal positivity, and chromogranin, synaptophysin and cytokeratin were negative. The diagnosis of PNET was made. The patient was started on a multidrug chemotherapy regimen. Although follow-up scans were done early after treatment the response to chemotherapy was poor with hardly any reduction in tumour size after 3 months.

Discussion

PNETs are very rarely seen in the black population³ and comprise 4% of soft-tissue tumours.² Peripheral primitive neuroectodermal tumours (pPNET) is a group of poorly differentiated small round blue cell tumours from neural crest origin.^{1-4,6} PNET and ES are classified together into the Ewing family of tumours,^{2,6} which is comprised of tumours with poorly differentiated small round cells.^{2,6} PNET is distinguished by neural differentiation,² seen as Homer-Wright rosettes^{4,7} or Flexner-Wintersteiner rosettes.^{4,7} Immunohistochemistry is used to detect expression of at least 2 of the following neural antigens in order to make the diagnosis of PNET, namely neuron-specific enolase (NSE), protein S100, Leucine 7, and neurofilaments protein, chromogranin A or synaptophysin.^{6,7} A highly specific antibody, CD99 (MIC2), can be found in both PNET and ES with immunostaining.^{5,7} PNET is seen mostly in the thoracopulmonary region and is also known as Askins tumour when it is in the chest wall.^{2,3,6,8} Other sites are the kidney, retroperitoneum/paraspinal, head and neck region.² Uncommon sites are the uterus, ovary, bladder, testis, pancreas, parotid, skin, subcutaneous tissues, lung, adrenal glands, dura mater and small bowel.²

These tumours are rapid-growing masses but regional lymphadenopathy and metastases are uncommon. When they metastasise they tend to go to bone and lungs^{1-3,6,8} more often. The next most common sites documented are bone marrow,^{2,6} lymphnodes¹ and brain.⁸ Metastases have also been described in the liver,² mediastinum³ and chest wall.³

Imaging modalities used are CT, MRI and nuclear medicine. CT and MRI are done to demonstrate the size and extent of the lesion.² Soft tissue invasion can be demonstrated well on both modalities.⁸ CT showed soft tissue invasion just as well as MRI in several cases⁸ and demonstrated bony erosion and lung metastases better than MRI,⁸ but MRI is the best modality to demonstrate spinal cord involvement. In our case

the diaphragm was demonstrated more clearly on the MRI sequences and infiltration of the diaphragm could not be excluded on CT. Another important role of CT or MRI is to determine resectability and to detect metastases.^{2,3} They are also used in follow up to determine response to treatment² and to detect recurrences. TC 99m-MDP (bone scan) is of value in detecting distant bony metastases⁸ and 8 F-fluoro-2-deoxy-glucose (FDG)-positron emission tomography (PET) scan can be used to detect recurrence of intraspinal PNET.^{1,9}

CT

The CT picture is usually of heterogeneous soft tissue density.^{2,3} The mass can be isodense or slightly hypodense to muscle³ and larger tumours commonly have hypodense necrotic/cystic areas.^{2,3,6,8} Post-contrast enhancement is mostly inhomogeneous.^{3,6,8} Calcifications are seen in less than 10% of cases, but could be faint and speckled or stippled.^{6,8} Haemorrhage can be seen as a hyperdense area in the mass if present. Regional lymphadenopathy is rarely seen but chest wall invasion is more common and would be evidenced by pleural effusion, bony destruction, tumour nodules in the muscles and abnormal enhancement of the chest wall.⁸

MRI

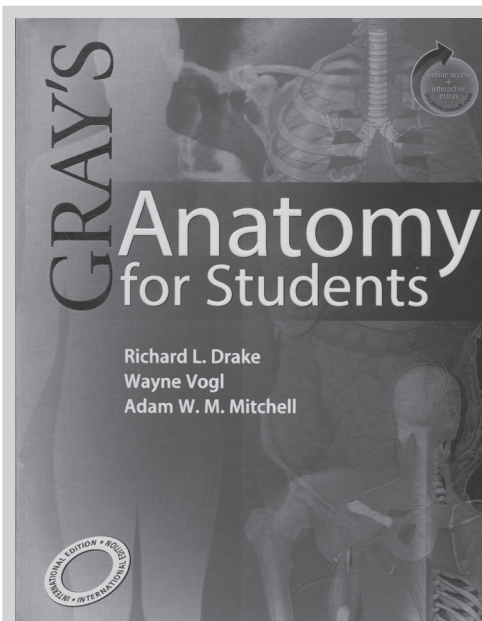
MRI T1-weighted images would show a mass isointense or slightly hyperintense to muscle^{2,3,5,6,8} with low-intensity areas correlating to cystic/necrotic areas in the tumour and hyperintense areas correlating to haemorrhage. Post-gadolinium enhancement may be seen uniformly or inhomogeneously.^{2,3,5,6,8} Heterogeneous high signal intensity is typically seen on T2-weighted images;^{2,3,5,6,8} this sequence usually demonstrates

the cystic components best. A STIR sequence would also demonstrate a heterogeneous high signal intensity mass.^{3,6}

Conclusion

PNETs are aggressive neoplasms and should therefore be diagnosed accurately and as early as possible. The distinction between PNET and ES cannot be made radiologically and could even be difficult on histological examination. Neural differentiation, immunostaining and immunohistochemistry can help to distinguish these tumours. Unfortunately a standard therapy does not exist yet and patients are offered a combination of surgery, chemo- and radiotherapy. Prognosis depends on the location of the tumour but PNET has a generally poor prognosis. Alternative treatment should be investigated further.

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