Clinical Radiology 69 (2014) 179–185

Contents lists available at SciVerse ScienceDirect

Clinical Radiology

journal homepage: www.clinicalradiologyonline.net

Imaging findings of the spinal peripheral Ewing's sarcoma family of tumours

W.-Y. Huang [a,c], W.-L. Tan [a,c], D.-Y. Geng [a,*], J. Zhang [a,*], G. Wu [b], B.-Y. Zhang [a], Y.-X. Li [a], B. Yin [a]

[a] Department of Radiology, Huashan Hospital, Fudan University, Shanghai 200041, China
[b] Department of Radiation Oncology, Shanghai Cancer Center, Fudan University, Shanghai, China

ARTICLE INFORMATION

Article history:
Received 1 May 2013
Received in revised form
10 August 2013
Accepted 12 September 2013

AIM: To present the neuroradiological and clinical characteristics of Ewing's sarcoma family of tumours (ESFTs) and to increase awareness of this neoplasm.

MATERIALS AND METHODS: The magnetic resonance imaging (MRI) features and clinical presentations of seven patients with pathologically documented ESFTs were retrospectively analysed. The tumour location, morphological features, signal intensity, contrast enhancement characteristics, involvement of the paraspinal soft tissues, and adjacent bony structures were assessed.

RESULTS: Most of the ESFTs in young adults were well-circumscribed. The present study demonstrated that ESFTs often have a hypo- or iso-intense signal on T1-weighted imaging and an iso-intense signal on T2-weighted imaging. Spinal ESFTs tended to present homogeneous signal intensity and diffuse enhancement. ESFTs are more likely to occur in the thoracic spine and later to infiltrate into the paraspinal area or vertebral bone. A broad dural attachment is another common feature in the cases presented here.

CONCLUSIONS: ESFT is a rare neoplasm that can have significant overlap in imaging appearance compared with other spinal neoplasms. A well-demarcated extradural mass invading the paraspinal soft or vertebral bones, with iso-intense on T2 weighted imaging and homogeneous enhancement could facilitate the diagnosis of spinal ESFT.

Introduction

The term primitive neuroectodermal tumour (PNET) was first used to describe tumours composed of small, round and undifferentiated cells resembling germinal or matrix cells of the embryonic neural tube that arose in the central nervous systems (CNS) of children and young adults.[1] When PNETs arise in the brain or spinal cord, they are called central PNET (cPNET). The term peripheral PNET has been applied to non-CNS tumours of the soft tissue, bone, and nerve, which have the morphological attributes of the germinal neuroepithelium. Ewing sarcoma (ES) is a highly malignant bone tumour composed of uniform round small cells. Later, malignant soft-tissue tumours, morphologically indistinguishable from ES, have been reported and termed extraskeletal Ewing sarcoma. According to Bruckner et al.[2], ES and pPNET are histologically small round blue cell tumours. The majority of cases share a cytogenetic translocation t(11;22)(q24;q12) with occasional variations and a characteristic immunohistochemical staining profile. Both tumours show different degrees of neuroectodermal differentiation. ES tends to be poorly differentiated, whereas pPNET most often exhibits definite neuroectodermal differentiation. Although

* Guarantor and correspondent: D.-Y. Geng and J. Zhang, Department of Radiology, Huashan Hospital, Fudan University, Shanghai, China. Tel.: +86 21 5288 8350; fax: +86 21 6248 9191.
E-mail addresses: gengdaoying2245@163.com (D.-Y. Geng), zhangjun8288@sina.cn (J. Zhang).
[c] Both authors contributed equally to this work.

0009-9260/$ – see front matter © 2013 The Royal College of Radiologists. Published by Elsevier Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.crad.2013.09.010

once viewed as distinct entities, ES, Askin's tumour (arising from the chest wall), and pPNET are now all considered members of the Ewing's sarcoma family of tumours (ESFT). The histopathological diagnosis of ESFT was based on the current histological criteria defined by the World Health Organization (WHO).[3] ESFTs are most commonly developed in the second decade of life and show a slight male predominance. The most frequently reported sites of occurrence are the chest wall, lower extremities, trunk, kidney, and orbit.[4,5] However, spinal ESFTs are an exceedingly rare entity. To the authors' knowledge, fewer than 80 cases of spinal ESFT have been reported in the English literature to date,[6,7] most of which focused on the histogenetic, pathological, and clinical features, treatment and prognosis of ESFTs.[8] Only a small number of sporadic case reports have described the imaging findings of ESFTs.[9,10] The purpose of the present study was to analyse the magnetic resonance imaging (MRI) characteristics of spinal ESFT.

Materials and methods

Institutional review board approval was obtained, and the requirement for patient consent was waived for this retrospective study. Seven patients with surgically and histologically proven ESFT from January 2005 to December 2012 were reviewed.

The MRI images were acquired using a 1.5 T MRI unit (Sigma Excite HD Twinspeed; GE Medical Systems, Milwaukee, WI, USA) or a 3 T MRI unit (Signa Excite GEMSOC01; GE Medical Systems) with a synergy spine coil. Sagittal and axial T1-weighted [600 ms repetition time (TR)/9.4 ms echo time (TE), spin-echo (SE) images] and sagittal T2-weighted (3200 ms TR/112 ms TE SE images) images were captured for unenhanced MRI, with a field of view of 275 mm × 325 mm for sagittal imaging and of 245 mm × 245 mm for axial imaging, an image matrix of 256 × 128 or 384 × 512, and a section thickness of 3 mm with a 0.3 mm gap. Contrast-enhanced sagittal and axial, T1-weighted, SE MR images were obtained after the administration of gadolinium diethylenetriamine pentaacetic acid [Gd-DTPA, (0.1 mmol/kg body weight) Magnevist, Berlex Laboratories, Berlin, Germany].

All images were evaluated by three experienced radiologists who were blinded to the histopathological diagnosis. The final assessment was reached by consensus. The images were specifically evaluated for lesion location, size, shape, margin, signal intensity, characteristics of enhancement, presence of paraspinal soft-tissue mass, and involvement of vertebral bones. In addition, clinical data, such as age, sex, symptoms, duration of symptoms, histopathological features, and treatment, were reviewed.

Results

Epidemiology and clinical presentations

The mean age at presentation was 28 years, ranging from 6–63 years. All seven of these patients had no significant medical or other surgical histories (Table 1). Patients presented with a variety of neurological symptoms and/or local pain. Symptom duration varied from 3 days to 6 months with a median of 2 months. Neurological examinations mainly revealed a bilateral weakness of the upper or lower extremities (Table 2). No other abnormal results of the laboratory examination were found. All patients underwent preoperative MRI examinations. Negative results of the brain MRI excluded the possibility of cPNET involving the spine. Work-ups for metastases were negative. Five patients underwent total surgical resection of the tumours and two patients underwent partial resections. Five patients received chemotherapy plus 30–50 Gy radiation therapy after surgery, while two very young patients received chemotherapy alone due to the age. All patients were followed-up from 3–6 months. Local recurrence was found in two partial resection patients after surgery.

Histopathological features

All patients had tumours that were histopathologically composed of monotonous small round undifferentiated cells with high nuclear/cytoplasmic ratios and finely dispersed chromatin. Homer–Wright rosettes were observed in all lesions. Immunohistochemistry examinations revealed consistent positive staining for CD99 in the tumour cells of all cases.

Imaging findings

The neuroradiological findings of ESFTs are summarized in Table 1. Of seven patients, six had extradural lesions that tended to involve paraspinal tissue (Fig 1d–e) or vertebral bone (Figs 1 and 2c) which were dumbbell-shaped

Table 1
Magnetic resonance imaging findings in seven patients with Ewing's sarcoma family tumours.

Case no.	Gender	Age (years)	Level	Tumour location	Margin	T1-weighted imaging	T2-weighted imaging	Enhancement	Signal feature	Paraspinal involvement	Bone involvement
1	M	16	T7–9	Extradural	Well-defined	Iso	Iso	Slightly	Homogeneous	Yes	–
2	M	30	T12–L1	Extradural	Ill-defined	Hypo	Hyper	Obvious	Heterogeneous	Yes	+
3	M	48	C7–T1	Intradural extramedullary	Ill-defined	Iso	Iso	Obvious	Homogeneous	Yes	+
4	F	63	T6–7	Extradural	Well-defined	Hypo	Hyper	Moderate	Homogeneous	Yes	+
5	M	6	T7–9	Extradural	Well-defined	Iso	Hyper	Slightly	Homogeneous	No	+
6	M	23	L4–S1	Extradural	Well-defined	Iso	Iso	Moderate	Homogeneous	Yes	+
7	F	16	T11–L1	Extradural	Well-defined	Hypo	Hyper	–	Heterogeneous	No	–