111 年奇美醫院胸腔內科臨床病例討論會

日期：中華民國 111 年 11 月 01 日(星期二)

時間及地點：16:00-17:00

課程活動題目：Askin tumor in CxR

主講人:謝俊民主任

主辦單位：奇美醫院胸腔內科

課程地點:10 樓討論室

教育積分：台灣胸腔暨重症加護醫學會

參加對象：主辦單位所屬院內醫師

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摘要:

Primitive neuroectodermal tumors (PNETs) are rare **undifferentiated sarcomas** that are believed to develop from migrating embryonal cells of the neural crest, and have been considered primarily a **pediatric tumor.**
PNET of the chest wall was first described as a specific entity by Askin et al[1] in 1979.

The purpose of this presentation is to describe an unusual case of PNET of the chest wall occurring in a 31-year-old male and to analyze the radiological features of this rare disorder .

CASE PRESENTATION：

A 31-year-old male without significant past medical history presented to the emergency department at our institution with a 1 week history of fevers and right-sided pleuritic chest pain. Additional history revealed night sweats over the previous month, and a 10-pound weight loss over the two preceding months.
Physical exam was unrevealing with normal lung exam and no adenopathy or palpable chest wall mass appreciated. Chest radiograph revealed a 2cm parietal-based mass in the lateral side of right hemi thorax, at the level of the middle arch of the third right rib (fig 1).
The patient left the emergency department after a symptomatic treatment with an appointment one week later in the out patient department for further exploration by the medical specialist.
The patient was not present at his appointment but 4 months later came again to the emergency department for dorsal and low back pain. The initial radiological examination (chest and dorsolumbar spine) demonstrates a 7 cm lobulated parietal-based opacity centered on the middle arch of the third right rib with cortical rupture and bone destruction

of the affected area of the rib (fig 2 and 3).
Subsequent thoraco abdomino pelvic contrast-enhanced computed tomography (CT) revealed an 8 x 7 centimeter right lateral chest wall mass with expansion, destruction and cortical spread out of the middle arch of the right third rib. This parietal mass was spontaneously heterogeneous containing solid and necrotic areas and demonstrates intense contrast uptake in the solid portion after injection with better delineation of the necrotic areas (fig 4 and 5).
A small (0.5 cm) sub pleural nodule was also noted in the left lower lobe (fig 6).
A second lesion was noted at L5 vertebral body with destruction of the postero superior aspect of the vertebral body and extension to the epidural space leading to mild compression of the spinal cord (fig 7).

The left iliac bone showed also a 0.5 cm focal lytic area located at the iliac wing without expansion of the cortex.
Ultrasound-guided trans-thoracic fine needle aspirate and core biopsies were obtained, revealing small, uniform round cells with pseudorosette formation. Immunohistochemical staining was consistent with PNET.
Given the pathologic findings, the diagnosis of PNET was made.
The patient currently is undergoing combination therapy with 3 cycles of chemotherapy with cyclophosphamide, vincristine, and adriamycin.

DISCUSSION:

Primitive neuroectodermal tumors of the chest wall, also called "Askin tumors" are rare, highly malignant tumors that have traditionally been felt to occur only in the pediatric or young adult population. These tumors are often very large at the time of diagnosis. The most common manifestation of these tumors includes pain from mass effect, often of several months duration. Additional symptoms include progressive shortness of breath and dyspnea, fever, night sweats, and weight loss [3,4] . For our patient the age was unusual (36 years) and the clinical manifestation was right chest wall pain and fever and weight loss with 3 months later low back pain due to the secondary location in the lumbar spine and subsequent epidural invasion.
Radiographic studies are non-specific, but most often reveal a chest wall mass with local rib destruction and extension into the thoracic cavity.

CT imaging is helpful to delineate any bony involvement.
Tumors that are adjacent to critical neurological structures require immediate MRI scanning and consideration of emergent radiation therapy to prevent nerve damage.
Magnetic resonance imaging often allows more accurate delineation and localization of the tumor and is helpful for determining the presence and extent of tumor invasion and for tissue characterization[5,6] .
Metastatic evaluation includes chest CT scanning and radioisotope bone scanning[6] .
Metastases to both pulmonary and extra-pulmonary sites are common. In our case at least

three secondary locations were noted in the left lung, in the left iliac bone and in the lumbar spine with epidural invasion [4,6] .

The typical histological features include uniform sheets of round, blue cells with round nuclei, sparse cytoplasm, a high mitotic index, and Homer-Wright pseudo-rosette formation, Immunohistochemical staining most commonly reveals staining for neuron-specific enolase (NSE) and vimentine, both of which were positive in this case. PNETs also have a cytogenetic translocation, t (11; 22)(q24; q12) that is shared with Ewing's sarcoma, and occurs in approximately 90% of the Ewing's--PNETs that can be evaluated, suggesting a common origin [2] . This genetic abnormality was not checked in this patient.
Aside the possibility of askin tumor, the differential diagnosis in case of aggressive rapidly growing chest wall mass could be an Ewing sarcoma a lymphoma, Osteosarcoma, Fibro sarcoma, Reticulum cell sarcoma, Histiocytosis X, Bone metastases, Sub acute or chronic osteomyelitis.
This particular case is unusual in that it is only the 8th reported case of a PNET of the chest wall ("Askin tumor") in a patient over 30 years of age. Additionally, this patient's presentation was unusual for the short duration of pleuritic chest pain prior to diagnosis and also the appearance of low back pain 3 months later that warned us for a secondary metastatic lesion.

CONCLUSIONS:
There are no specific radiographic characteristics for Askin tumor that may be helpful in distinguishing this disorder from other small round cell tumors. Anatomopathological examination with inmunological test remains necessary to make a correct diagnosis.
The traditional differential diagnosis of a large chest wall mass in an adult has included lymphoma, sarcoma, metastatic lesions and benign lesions such as eosinophilic granuloma. However, this ease reiterates the need to consider primitive neuroectodermal tumor of the chest wall ("Askin tumor") in a patient of any age who presents with a chest wall mass.

Reference(s):

1 Askin FB, Rosai J, Sibley RK, et al. Malignant small-cell tumor of the thoraco-pulmonary region in childhood. Cancer 1979; 43:2438-2451
2 U. Tateishi, G. W. Gladish, M. Kusumoto, T. Hasegawa, R. Yokoyama, R. Tsuchiya, and N. Moriyama
Chest Wall Tumors: Radiologic Findings and Pathologic Correlation: Part 2. Malignant Tumors
RadioGraphics, November 1, 2003; 23(6): 1491 - 1508.
3 G. W. Gladish, B. M. Sabloff, R. F. Munden, M. T. Truong, J. J. Erasmus, and M. H. Chasen Primary Thoracic Sarcomas
RadioGraphics, May 1, 2002; 22(3): 621 - 637.
4 Verrill MW, Judson IR, Harmer CL: Ewing's sarcoma and primitive neuroectodermal tumor in adults: are they different from Ewing's sarcoma and primitive neuroectodermal tumor in children? J Clin Oncol 1997 Jul; 15(7): 2611-21

5 Triche TJ. Pathology of pediatric malignancies. In: Pizzo PA, Poplack ED, eds. Principles of practice of pediatric oncology. 2nd ed. Philadelphia: J.B. Lippincott, 1993:115-152
6 Stephenson CF, Bridge JA, Sandberg AA. Cytogenetic and pathologic aspects of Ewing's sarcoma and neuroectodermal tumors. Hum Pathol 1992; 23:1270-1277.