

Case report

POEMS syndrome: unusual radiographic, scintigraphic and CT features

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Abstract. POEMS syndrome is a multisystemic disorder related to a plasma cell dyscrasia. Radiologically, this syndrome is characterized by sclerotic focal bone lesions with a normal radionuclide bone scan. We report a case of POEMS syndrome with an expansile lytic lesion in the sternum showing periosteal reaction and soft tissue mass, which revealed locally increased uptake of radiotracer in bone scintigraphy. These unusual findings and the differential diagnosis are discussed.

Key words: POEMS syndrome – Plasmacytoma – CT – Scintigraphy

Introduction

In 1980 Bardwick proposed the acronym POEMS to describe a syndrome characterized by polyneuropathy, organomegaly, endocrinopathy, M-protein and skin changes [1]. The clue in the pathogeny is a plasma cell dyscrasia that originates the M-protein. The other systemic manifestations are due to obscure mechanisms probably correlated with this dyscrasia. Skeletal radiologic findings are present in nearly 95 % of patients and differ from the lytic lesions of classic myeloma in their sclerotic appearance [2]. Purely lytic lesions in POEMS are uncommon. Bone scintigraphy is usually negative with few exceptions. Our patient presented a sternal osteolytic lesion with an aggressive radiographic appearance – sunburst periosteal reaction and soft tissue mass – and an increased radionuclide uptake with ^{99m}Tc-methylene diphosphonate scintigraphy.

Case report

A 50-year-old man who had a history of chronic sensorimotor polyneuropathy of unknown etiology, was re-

ferred with a 2-year history of painless swelling of the sternal manubrium. He could not remember any antecedent symptoms of trauma or infection. Questioning revealed impotence and weight loss over the previous few months.

Examination on admission showed a painless palpable mass over the sternal manubrium, tender hepatomegaly, axillary lymphadenopathy, bilateral papilledema, multiple cutaneous angiomas, gynecomastia and testicular atrophy. Laboratory data were unremarkable except for elevated hemoglobin, hematocrit and total serum protein. Serum electrophoresis disclosed a spike of IgG lambda type; Bence-Jones proteinuria was ruled out. Thyroid stimulating hormone (TSH) level was increased, while T₃ and T₄ were normal. Cerebrospinal fluid analysis revealed elevated protein levels. Bone marrow aspiration and biopsy were normal.

On a radiologic survey there was a sternal expansile lytic lesion with an ill-defined spiculated margin (Fig.1). Increased bone uptake was found on ^{99m}Tc-MDP scintigraphy (Fig.2).

CT disclosed a lytic lesion involving the sternal manubrium with few thin intralesional trabeculae. This lesion showed partial cortical destruction with inhomogeneous sclerosis of the remaining cortex. There were also a periosteal 'sunburst' new bone formation (Fig.3) and a bulky anterior soft tissue mass protruding into the anterior thoracic wall. CT-guided fine-needle biopsy was performed twice, but yielded insufficient material for diagnosis.

At surgery, an 'en bloc' resection of the sternum was performed. Pathologic examination of the surgical specimen demonstrated a monoclonal IgG lambda plasmacytoma.

Discussion

The main clinical features of POEMS syndrome are plasma cell dyscrasia which produces an M-protein, progressive motor or sensorimotor polyneuropathy, orga-

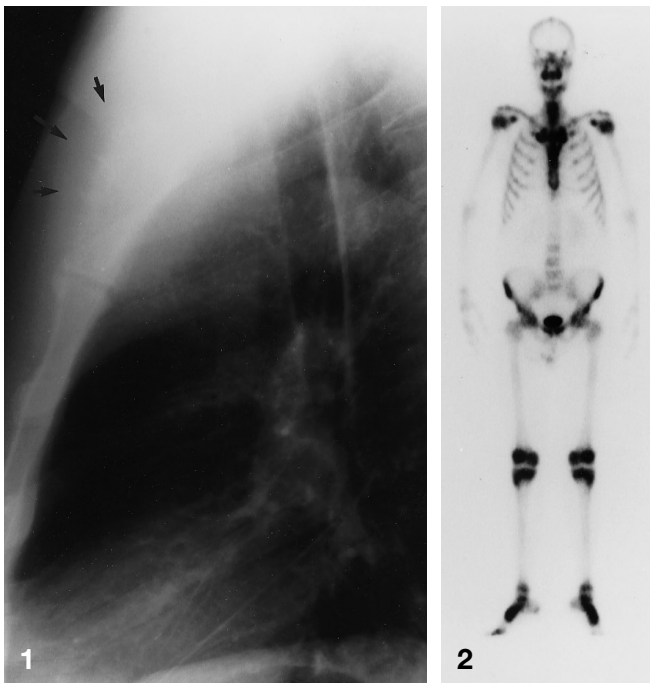


Fig. 1. Plain radiograph shows a lytic mass in the sternum (*arrows*). Note the spiculated anterior border of the lesion

Fig. 2. Bone scan demonstrates increased uptake at the sternal manubrium



Fig. 3. CT of the sternum shows an osteolytic lesion with marked sclerosis and disruption of the cortex. There was a 'sunburst' periosteal new bone formation (*arrows*) and an anterior large soft tissue mass

nomegaly (hepatosplenomegaly, lymphadenopathy), endocrinopathy (hypothyroidism, gynecomastia, impotence, diabetes mellitus, hyperthyroidism and suprarenal insufficiency), skin changes (hyperpigmentation, hypertrichosis, skin thickening and angiomas), anasarca, papilledema and bone lesions.

POEMS differs from classic myeloma in a certain number of features. This syndrome affects younger patients, there is a predominance of lambda light chains, a lower level of M-protein in blood and a lesser degree of bone marrow infiltration by plasma cells [1, 3]. In multi-

ple myeloma the most frequent clinical manifestation is bone pain [4], which is unusual in POEMS syndrome [2, 3] although skeletal abnormalities are present in 54–95 % of patients [1, 2, 6]. There are two types of skeletal manifestations: proliferative changes and focal bone lesions. Osseous proliferation, especially at the posterior elements of the lumbar spine, has been considered virtually pathognomonic for POEMS syndrome [1, 6, 7].

Focal lesions can be solitary or multiple, and they are distributed primarily in the axial and proximal appendicular skeleton. Sternal location is not frequent. Frequently a diagnosis of plasmacytoma is suggested at histological study [5]. Radiologically, these lesions are usually sclerotic, or of mixed lytic-sclerotic appearance, consisting of a lucent lesion with sclerotic rim. The differential diagnosis of these sclerotic bone lesions includes metastases, tuberous sclerosis, mastocytosis, cystic angiomas and Hodgkin disease [1, 6, 7].

In our case, radiography and CT disclosed an expansile osteolytic lesion with very aggressive features: cortical destruction, soft tissue mass, and periosteal new bone formation.

In POEMS syndrome, purely lytic lesions are infrequent [1, 6]. In previously published reports of POEMS syndrome with lytic lesions little attention has been paid to its radiologic characteristics. Most of these cases are described only as osteolytic lesions, without further characterization. Bessler [8] reported a case of POEMS syndrome with two lytic expansile lesions in flat bones showing periosteal reaction. As in our patient, the costal lesion of this case contained sclerotic intratumoral trabeculae; otherwise, the periosteal reaction was of solid type. Classic myeloma and solitary plasmacytoma involving flat bones, such as the sternum or the ribs, can also appear as expansile lytic lesions [4], but periosteal reaction is unusual in untreated myeloma and plasmacytoma [4].

In our case, the clinical setting suggested the diagnosis of sternal plasmacytoma in POEMS syndrome. Furthermore, the 2-year history of painless swelling reinforced this presumed diagnosis. Nevertheless, the aggressive appearance of the lesion made it imperative to consider osteosarcoma or metastases as more probable radiologic diagnoses.

Skeletal scintigraphy is usually negative in POEMS syndrome. Only one case of POEMS syndrome with a positive bone scan has previously been reported [8]. In the patient of Bessler [8], the lytic lesions showed radionuclide uptake and the authors suggested that this accumulation of radionuclide was due to cortical expansion, periostitis and sclerotic reaction of the surrounding bone. In our case, the increased radionuclide uptake was more than probably explained by cortical sclerosis and periosteal bone formation.

In summary, we report a case of POEMS syndrome presenting unusual radiologic and scintigraphic features. While the sclerotic aspect of bone lesions and its inactivity in bone scintigraphy have been considered the main radiologic differential aspects of this entity, our case shows an aggressive osteolytic lesion with positive scintigraphy.

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Book review

European
Radiology

Ansell, G., Bettmann, M. A., Kaufman, J. A., Wilkins, R. A.: Complications in Diagnostic Imaging and Interventional Radiology, 3rd edn. Oxford: Blackwell Science 1996. 720 pp., 253 figs., (ISBN 0-86542-243-5), £ 160.00.

Radiology has changed dramatically over the last 25 years. In the early 1970s diagnostic imaging studies using X-rays were the only procedures performed by radiologists. Now a variety of imaging procedures are available including ultrasound, computed tomography, digital subtraction angiography, and MRI. Moreover, interventional radiology has opened new horizons for minimally invasive vascular and nonvascular diagnostic and therapeutic procedures. Thus, the radiologist at the end of this century needs not only a profound knowledge of the potential hazards and complications of radiation, but must also be aware of the potential risks of any type of interventional, imaging-guided procedure, the appropriate use of pharmaceuticals for interventional procedures or the treatment of any potential complications, and the hazards associated with a strong static magnetic field or a fast and strong gradient field.

The purpose of this book is to address all these different topics and to provide a comprehensive overview. The book is divided into five parts: patient care, technical considerations, intravascular contrast media, organ systems, and risk identification and management.

The part on patient care discusses a variety of topics including a general overview of anesthesia, the use of drugs in the imaging department, a brief review of cardiac physiology, infectious complications in angiography and percutaneous biopsies, and the care of the pregnant and nursing patient. The chapter on drugs is particularly well organized and discusses systematically the indications, dosages, side-effects and contraindications for the most important drugs which may be required in diagnostic and interventional procedures. In contrast, the chapter on radiation exposure in interventional radiology is not so helpful. More tables and figures displaying data about exposure rates for the patient and the doctor would have been more helpful.

The part dealing with technical considerations covers a wide variety of different fields: nuclear medicine, MRI, lasers, diagnostic ultrasound, and electrical and mechanical hazards which may potentially occur in an imaging department. The MRI chapter contains a large table summarizing the bioeffects of MRI. However, it would have been more helpful for the reader who is not familiar with MRI to extend the list of metallic implants, materials and foreign bodies which might be a risk for a patient undergoing MRI-in-

formation which is well known from original articles and textbooks by the same authors.

The third part discusses in detail mechanisms, manifestation and treatment of the complications of intravascular contrast media. However, when reading this chapter one is struck by the absence of practical advice on how to prepare a patient with impaired renal function for an imaging study using an intravenous contrast agent.

The largest part of this book is dedicated to a discussion covering more than 300 pages of the potential risks of any type of interventional imaging-guided procedure, including inpatient diagnostic angiography, outpatient angiography, arterial and venous stent placement, TIPS, embolization, IVC filter placement, regional fibrinolytic therapy, biliary interventions, genitourinary tract interventions, interventional neuroradiology, and percutaneous procedures such as fine needle aspiration biopsy of thoracic lesions, abscess drainage, breast biopsies and orthopedic procedures. Moreover, there are chapters discussing complications of endoscopy and procedures in gynecology. The final chapter is dedicated to interventional procedures in the pediatric patient population.

When reading the different chapters it becomes obvious that sometimes only the surface of a problem has been scratched. One is missing more technical and practical advice that is necessary to overcome certain problems. Moreover, the image quality is sometimes poor, as in the chapter on venous vascular stenting, and occasional images illustrating complications are simply missing, as in the chapter discussing complications in the gastrointestinal tract. The chapter on regional fibrinolysis is particularly worthwhile, as it offers a comprehensive review of the literature and gives the reader good practical advice.

In the fifth part of the book some general considerations and some detailed considerations dealing with medicolegal problems in the United States and the United Kingdom are addressed. They may be of minor interest to a radiologist outside these countries.

In summary, this book offers a fairly complete, general overview of the potential complications of diagnostic imaging and interventional radiology at a moderate price. It can be recommended for the resident who is starting training in interventional radiology or other subspecialty of diagnostic radiology or to the general radiologist who feels a need for basic well-accepted information to present to patients in order to obtain informed consent. For all those who want to go into more depth, the relatively complete bibliography at the end of each chapter allows a fast orientation to and facilitates finding of the appropriate original articles.

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