

Extraosseous myeloma: imaging features

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Learning objectives

- To review the imaging findings of extraosseous myeloma.
- To stand out the importance of knowing this unusual myeloma manifestation.
- To know the differential diagnosis of this pathology.

Background

Multiple myeloma is a hematologic disorder characterized by a proliferation of clonal plasma cell of B-lymphocyte origin in the bone marrow. It accounts for approximately 1% of all malignant diseases and represents about 10% of hematologic malignancies in adult population. The peak age of onset is during the sixth and seventh decades, and it is more common in males than in females.

Extraosseous myeloma refers to the presence of myeloma deposits outside the skeletal system. Typical radiological manifestations of myeloma as diffuse osteopenia and osteolytic bone lesions are well recognized. However radiological diagnosis of an extramedullary myeloma is very rare.

Although classically extraosseous myeloma has been considered uncommon, according to autopsy studies, extraosseous spread was present in approximately 63.5% of myeloma patients. The lymph nodes, the pleura, the spleen and the liver are the organ most commonly involved, although it can affect any organ.

Extraosseous disease is usually associated with a more aggressive form of multiple myeloma.

We retrospectively reviewed the imaging studies of 6 patients with extraosseous myeloma in our hospital from January 2009 to September 2013. Four patients were men and two were women (age range, 30-82 years, mean age, 54 years). Extraskkeletal tumors were present in the lung, spleen, liver, subscapularis muscle, cavum, hypothalamus and pineal gland.

Findings and procedure details

There is no consensus in the imaging evaluation of patients diagnosed with multiple myeloma. Usually extraosseous myeloma is an incidental finding or a search oriented to the presence of symptoms.

The imaging features are not specific, so we have to consider the possibility of extraosseous myeloma in focal soft tissue masses in patients with known multiple myeloma.

HEPATIC

The hepatic involvement usually is diffuse and hepatomegaly may be seen. In the published case reports tumor nodules appear often hypoechoic or with a “target” appearance (isoechoic nodule with a hypoechoic rim) on ultrasound.

On CT the most common finding is the presence of focal lesions with a low attenuation that do not show calcification or significant contrast enhancement.

On MRI the myelomatous lesions are usually hyperintense on T1-weighted images (possibly due to the high protein content) with a high signal on T2-weighted images. After gadolinium, minimal enhancement has been observed within the lesions.

Because the imaging findings are nonspecific, in general the final diagnosis is confirmed by the histologic study.

In our hospital we observed 3 patients with hepatic involvement, one of them had multiple focal liver lesions varying in size from 0,5 to 4 cm, another patient had two lesions and one patient had a solitary focal

mass. All the nodules appeared hypodense without significant contrast enhancement.

PANCREATIC

Multiple autopsy studies have revealed that extraosseous involvement in the pancreas range between 4% and 17% of patients. On ultrasound, the pancreatic masses may manifest as unifocal or multifocal hypoechoic, heterogeneous lesions. Biliary obstruction may occur if myelomatous involvement is prominent within the pancreatic head.

In the literature they are usually described as hypervascular masses on Ct, thus they are often mistaken for neuroendocrine tumors.

We have observed a solitary head pancreatic mass in one of our patients. It was described as a hypodense lesion with mild dilatation of pancreatic duct and common bile duct.

SPLENIC

Splenic involvement is seen between 30% to 45% of patients on autopsy. Splenomegaly is often the only imaging finding. Splenic involvement is also more commonly diffuse than focal. Focal myeloma deposits, may be seen hypoechoic on ultrasound and hypodense on CT. It is usually seen in association with hepatic involvement.

In our hospital we observed a patient with a homogeneous splenomegaly with diffuse myelomatous involvement.

GASTROINTESTINAL

The small bowel is the most common site of gastrointestinal involvement by extraosseous myeloma, followed by the stomach and colon. The radiological findings include diffuse mural thickening, polyps and parietal or intraluminal masses.

LYMPH NODES

Myeloma involvement of the lymph nodes is typically seen as enlarged homogeneous nodal masses. Multiple nodal stations are

affected. It is essential to make a good differential diagnosis with the lymphoma as both pathologies show multiple nodal affectation with a low T2 signal on MRI.

GENITOURINARY SYSTEM

Renal involvement has been described as focal masses, diffuse involvement and perirenal nodules. Testicular involvement has also been described as a hypoechoic mass or diffuse enlargement.

Nor renal nor testicular mass were seen in our institution.

HEAD AND NECK

Diffuse infiltration of the leptomeninges is the most common site of central nervous system involvement. It is usually seen as diffuse enhancement resembling meningitis or as focal masses similar to meningiomas.

Extraosseous myeloma has a predilection for the nasal cavity. They may be seen as masses which are hypointense on T2-weighted images and enhance homogeneously.

We have observed a hypodense mass located in the cavum in a patient. In another patient two focal lesions were described, one located in the pineal gland and the other one in the hypothalamus. They appeared as hypointense mass on T1 and T2-weighted images with a homogeneous enhancement and vasogenic edema.

THORAX

Extraosseous myeloma in the thorax occurs most commonly in the form of pulmonary parenchymal nodules, masses, or interstitial infiltration.

In our case a lung parenchymal mass obstructing the intermediate bronchus was present in one patient with extensive mediastinal lymph nodes and hepatic involvement also.

MISCELLANEOUS SITES

Homogeneous soft-tissue nodules can be seen in subcutaneous tissues or within the muscles mimicking lymphoma, metastases, sarcoma or desmoid tumors.

On imaging, the myelomatous deposit usually presents as a well-defined subcutaneous nodule/mass. In our case we have a patient with multiple hypodense nodules on the subcutaneous tissue and a hyperdense nodule in the subscapularis muscle.

Conclusion

-Extraosseous myeloma can affect any part of the body and is associated with aggressive disease and young patients.

-Among the common imaging findings of extraosseous myeloma we can find homogeneous soft-tissue masses on CT and masses with a low density on T2-weighted images. However, the imaging features are not specific and we have to consider the possibility of extraosseous myeloma in focal masses in patients with known multiple myeloma. The biopsy is usually required to confirm diagnosis.

References

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Fig. 1: Axial CT (1: non-enhanced, 2: arterial phase contrast-enhanced, 3: portal phase contrast-enhanced) showing a solitary hypodense lesion in the left lobe of the liver without significant contrast enhancement in a patient with myeloma.

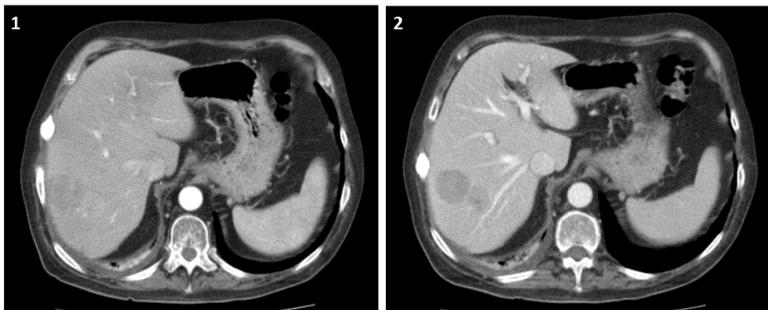


Fig. 2: Axial CT (1: arterial phase contrast-enhanced, 2: portal phase contrast-enhanced) showing two hypodense lesions in the right lobe of the liver without significant contrast enhancement in a patient with myeloma.



Fig. 3: Axial CT (1: non-enhanced, 2: arterial phase contrast-enhanced, 3: portal phase contrast-enhanced) showing multiple hypodense focal liver lesions varying in size from 0.5 to 4 cm in a patient with myeloma.

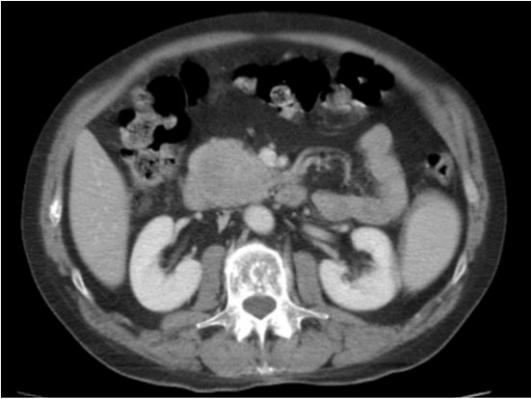


Fig. 4: Axial contrast-CT scan shows a hypodense head pancreatic mass with mild dilatation of pancreatic duct and common bile duct.

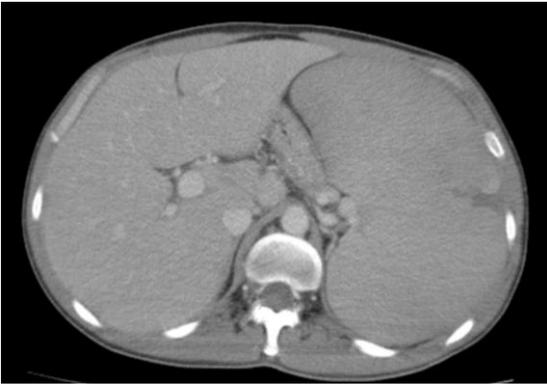


Fig. 5: Contrast-CT scan showed a homogeneous splenomegaly in patient with myeloma. Percutaneous biopsy revealed sheets of plasma cells.

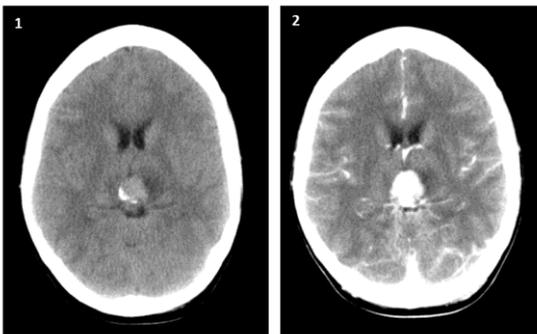


Fig. 6: 1: Axial non-enhanced CT scan. 2: Axial contrast-CT scan. The images show a hyperdense focal lesion in the pineal gland that enhances homogeneously after the administration of contrast in a patient with myeloma.

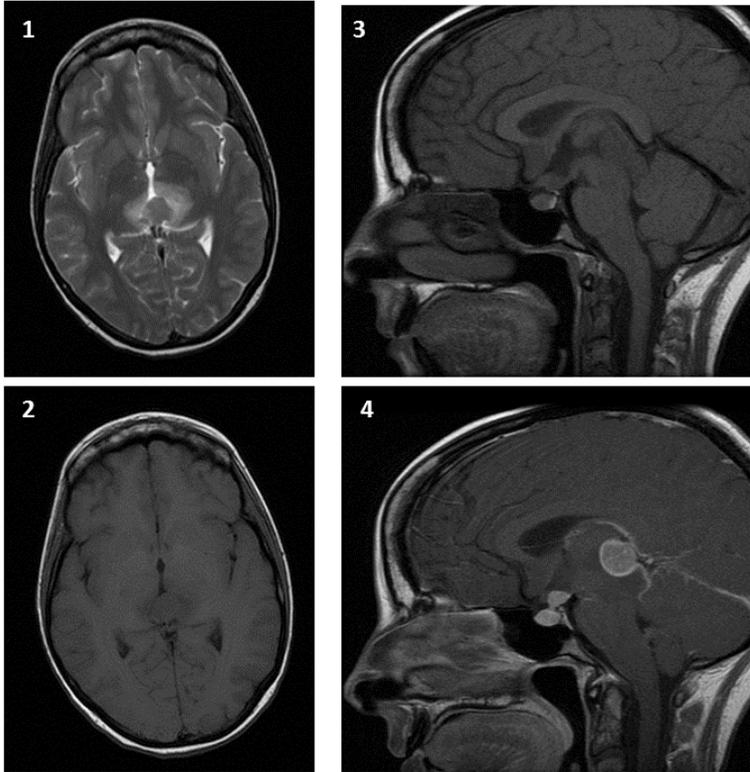


Fig. 7: 1: A T2-weighted axial MR image. 2:A T1-weighted axial MR image. 3: A T1-weighted sagittal MR image. 4: A contrast-enhanced T1-weighted MRI. They show a hypointense mass in the gland pineal and another one in the hypothalamus with a homogeneous enhancement and vasogenic edema surrounding the lesions.



Fig. 8: Axial contrast-CT scan shows a hypodense mass located in the cavum of a patient with myeloma.



Fig. 9: Axial contrast-CT scan shows a lung parenchymal mass obstructing the intermediate bronchus in one patient with extensive mediastinal lymph nodes and hepatic involvement also (not seen in this image)

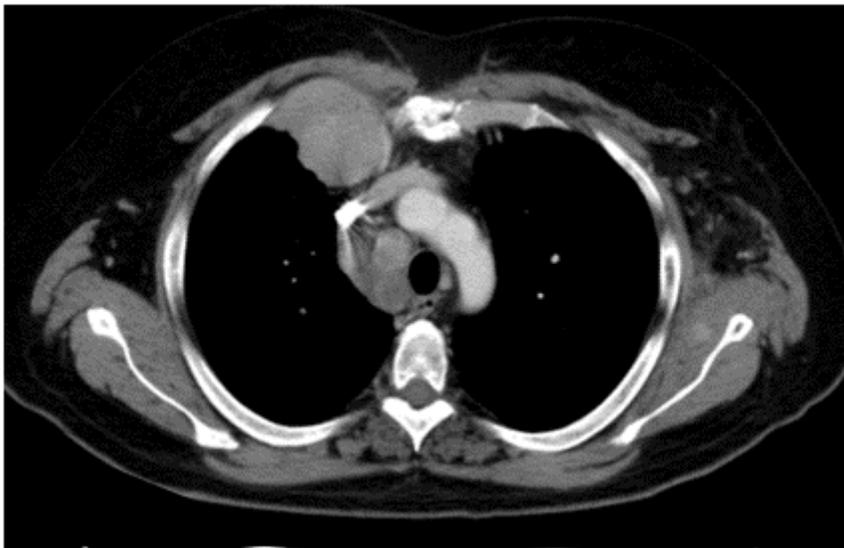


Fig. 10: Axial contrast-CT scan shows a right pericostal tumor mass caused by direct spread from adjacent bony lesion and a hyperdense nodule in the left subscapularis muscle.