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CASE REPORT

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Plasma cell myeloma with tumorous liver involvement, presenting as a carcinoma of unknown primary

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Abstract

Background: *A hepatic tumor mass by plasma cell myeloma is rare and is reported in 16% of patients with plasma cell myeloma. Unusual morphological variants of plasma cell myeloma in such a localisation may be a diagnostic challenge.*

Case presentation: *A 60-year-old woman with low-back pain had MRT and CT scan results showing osteo-destructive lesions on different sites and a tumor mass in the liver. Her clinical presentation was classified as a carcinoma of unknown primary. Elevated serum-levels of the immunoglobulin heavy chain IgA and the immunoglobulin light chain kappa gave rise for a bone marrow trephine biopsy, revealing infiltration by a plasma cell myeloma with typical morphology (Fig.1A). Though the histological specimen of the liver resembled a clear cell carcinoma, the immunophenotype revealed plasma cell myeloma.*

Conclusion: *In tumours with an extraordinary morphology and not conclusive immunohistochemical results, the use of plasma cell antibodies may lead to the correct diagnosis.*

Keywords: *plasma cell myeloma, clear cell carcinoma, liver*

Abbreviations: *WHO: World Health Organisation; CUP: Carcinoma of Unknown Primary.*

Background

Plasma cell myeloma is a rather common neoplasm accounting 10-15% of the haematopoietic neoplasms and 20% of deaths from haematological neoplasms. In the WHO classification, plasma cell myeloma [1,2] is defined as a bone marrow-based, multifocal neoplastic proliferation of plasma cells, usually associated with an M protein in serum and /or urine and evidence of organ damage related to the plasma cell neoplasm. The most common morphological variants of plasma cell myeloma show a plasmacytic, plasmablastic or pleomorphic appearance. However, there are reports of unusual morphological variants, which do not reveal a plasmacytic morphology [3-9]. An extra-medullary spread can occur, in fact, it has been reported to be present at the time of initial diagnosis in 7%-17% of patients and it appears during the course of the disease in 6%-20% of the patients [10]. During autopsy, a hepatic infiltration is described in up to 40% of the cases, most commonly as diffuse sinusoidal Infiltartes. Only 16% of the cases show a liver tumor mass [1]. Extra-medullary involvement is associated with aggressive disease progression and biological and histological features of poor prognosis such as blastic histology [10].

Case presentation

AA 60-year-old female patient with low-back pain lasting for about two months, had an MRT for diagnostic clarification. The MRT results showed osteo-destructive lesions of the lumbar spine, the ribs and the iliac bone. CT-Scan showed a tumour-mass of 3.7 cm in diameter in the left ventral thorax-wall, a 3 cm in diameter osteo-destructive lesion in L4 vertebral body and a tumour of 4 cm in diameter in the liver. A mammography was inconspicuous. The clinical picture was suspicious of a carcinoma of unknown primary. Further examination revealed elevated serum-levels of the immunoglobulin heavy chain IgA and the immunoglobulin light chain kappa. A bone marrow trephine biopsy was performed revealing infiltration by a plasma cell myeloma with typical morphology (Figure 1A) and expression of the immunoglobulin heavy chain IgA, the immunoglobulin light chain kappa and co-expression of CD56. The extent of bone marrow infiltration was 25% compared to the haematopoiesis. Because of the tumorous lesions found in CT-scan a liver biopsy was performed to exclude a secondary neoplasm. The morphology of the liver tumour resembled a clear cell carcinoma. However various immunohistochemical stains for carcinoma were negative. With the knowledge of the patient`s plasma cell myeloma, relevant plasma cell marker inclusive light and heavy chain antibodies were performed, leading to the correct diagnosis of a plasma cell myeloma. Despite extensive treatments, the disease progressed. A bone marrow biopsy taken 11 months after primary diagnosis showed neoplastic plasma cell infiltration, quantified by 98% compared to the haematopoiesis. The patient ultimately died of bone marrow failure.

Results

The histological examination of the liver tumour showed tumour cells with round and partially cleaved nuclei with small nucleoli and a clear broad cytoplasm (Figure 1B and 1C). Some tumour cells were polynucleated, some showed bizarre nuclei. The morphology resembled a clear cell carcinoma. Immunohistochemistry using

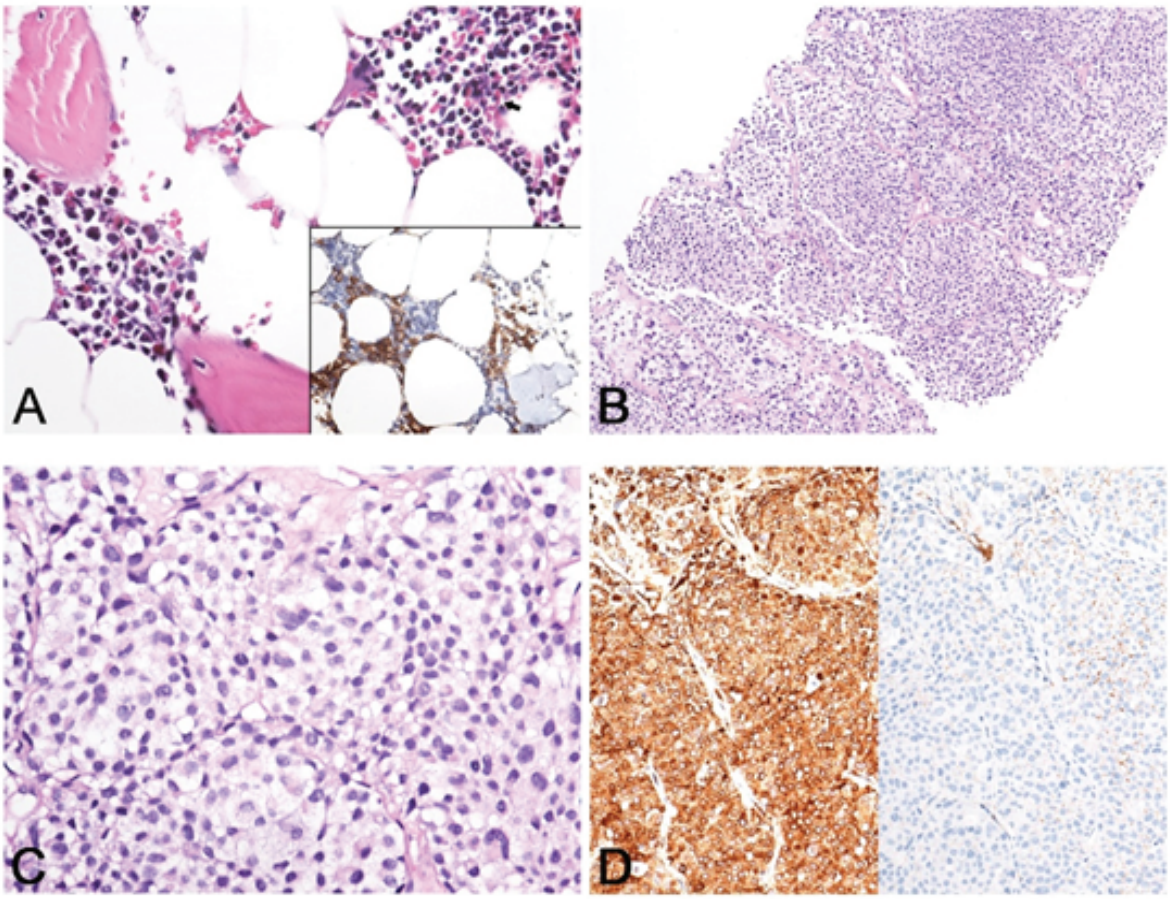

Figure 1: *(A): Bone marrow biopsy, haematoxylin eosin staining, (the arrow points to the plasmacytically differentiated infiltrate) (40x) with inset picture of CD38 immunohistochemistry (40x). (B): Liver biopsy, haematoxylin eosin staining (10x). (C): Liver biopsy, haematoxylin eosin staining (20x). (D): Liver biopsy, immunohistochemistry (20x) Kappa (left) and lambda (right).*

antibodies against epithelial tumour cells and malignant melanoma were negative. Since the diagnosis of plasma cell myeloma was known, immunohistochemistry (Figure 1D) with plasma cell markers was performed confirming a plasma cell myeloma in the liver. .

Discussion

The diagnosis of an extra-medullary plasmacytoma or an extra-medullary involvement of a plasma cell myeloma usually is not challenging. The patient described here, had a clinical picture which was initially interpreted as a carcinoma of unknown primary ("CUP"). The elevated serum levels of the Immunoglobulin heavy chain IgA and the Immunoglobulin light chain kappa led the clinicians to the diagnosis of a plasma cell myeloma. This neoplasm was confirmed by bone marrow biopsy. In the bone marrow, the neoplastic plasma cells showed the typical morphology in contrast to the liver tumour.

In cases with unusual histological morphology, the way to the correct diagnosis can be challenging. In this case, the differential diagnosis of a second neoplasm such as undifferentiated malignant tumour had to be considered. The positivity for CD56 could be misinterpreted as neuroendocrine differentiated carcinoma. The differential diagnosis should include malignant lymphoma, poorly differentiated adenocarcinoma, clear cell carcinoma, high-grade sarcoma and malignant melanoma.

Conclusion

A Liver tumor mass as an extra-medullary manifestation of a plasma cell myeloma is uncommon [11]. In this case, the tumor showed the appearance of a clear cell carcinoma, so its morphology did not correspond to the plasma cell myeloma diagnosed in the bone marrow. It is to mention, that without the knowledge of the plasma cell myeloma the diagnostic procedure would be more difficult. Although morphological variants of plasma cell myeloma are rare, a pathologist should be aware of their existence. Regardless of the location, in tumours with an extraordinary morphology and a not conclusive immunprofile the use of plasma cell antibodies may lead to the correct diagnosis.

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