Extradural Plasmacytoma of the Gallbladder with t(14;16) IGH/MAF - Case Report.

Jung Moon, MD, MMS, Kenneth N. Holder, MD
The University of Texas Health Science Center at San Antonio, TX

Background

Extradural plasmacytoma is a rare plasma cell neoplasm that can present with or without bone marrow involvement or other systemic characteristics of multiple myeloma. Primary gallbladder plasmacytoma is a rare type of extradural presentation with only 8 cases having been reported in literature; 2 with bone marrow involvement. We present a unique case of gallbladder plasmacytoma that was detected on ultrasound image guided biopsy and was found to be t(14;16) positive.

Methods

A 69-year-old woman presented for evaluation of bilateral knee pain and pathological fracture of the tibia. Imaging with MRI found enhancing intramedullary lesions within the proximal left tibia, and abdominal CT done during work-up found a lobulated enhancing mass measuring 7.9 x 7.1 x 7.7 cm arising from the gallbladder fundus, compressing hepatic segment VI. An ultrasound-guided core needle biopsy of the gallbladder mass was performed.

Results

Examination of the gallbladder biopsy specimen showed expansile sheets of atypical plasma cells expressing CD138, partial CD56, and lambda light chain by in-situ hybridization on immunohistochemistry and lacking CD3, CD20, EBER, S-100, pancytokeratin AE1/AE3, and kappa. There was no morphologic evidence of gallbladder or other normal structure in the biopsy; however, imaging studies demonstrate accurate placement of the core needle biopsy within the gallbladder mass. Interphase fluorescence in situ hybridization analysis (FISH) on paraffin-embedded tissue found the presence of t(14;16) IGH/MAF (ISCN : nuc ish(IGH, MAF)x3(IGH con MAF x 2)) with a gain of the IGH/MAF fusion gene. Subsequent to the diagnosis, serum protein electrophoresis and immunofixation electrophoresis demonstrated an IgG-lambda type monoclonal gammapathy with a M-protein of 0.6g/dL, and the free kappa/lambda ratio on serum was 0.002. The complete blood count, serum calcium, and creatinine were all within normal limits. A subsequent skeletal survey and CT/MRI showed multiple lytic/lucent lesions of the calvarium and throughout the thoracic spine and bilateral scapula, sacrum, pelvis and left femur concerning for diffuse osseous metastases. A posterior iliac crest bone marrow biopsy was performed and no abnormal plasma cell population was detected either by morphology or immunophenotyping. A repeat and bilateral bone marrow biopsy showed focal involvement on one side and a directed biopsy of a lytic lesion of the femur demonstrated involvement by plasma cell neoplasm with immunophenotype identical to the above (both performed after abstract submission). The patient was recently started on chemotherpay (cyclophosphamide, bortezomib, dexametasone) and zometa.

Conclusion

This case reports a ninth example of gallbladder plasmacytoma and is the first to report a cytogenetic abnormality detected by FISH. Various studies have suggested that t(14;16) positive plasma cell myeloma is associated with less frequent extradural involvement, IgG M-protein, high protein levels, CD20 positivity, CD56 negativity, hypercalcemia, leukocytosis, thrombocytopenia, a poor prognosis, and resistance to proteasome inhibitor (PI) therapy. In contrast, this case demonstrates a t(14;16) positive plasma cell neoplasm with extradural involvement and CD56 positivity. Initial radiographic studies in this case were suggestive of a gallbladder carcinoïd with metastases; however, results of the biopsy confirming a plasmacytoma demonstrate the need for careful clinical correlation and a broad differential when approaching rare gallbladder masses.

References


Table 1: Summary of the literature on the presentation and prognosis of extramedullary plasmacytomas.