An Unusual Cause of Isolated Fundal Gastric Varix

Fahad Chaudhary, MD
West Virginia University, Department of Medicine. Morgantown, WV

Hiren Vallabh, MD
West Virginia University, Section of Digestive Diseases. Morgantown, WV

Sardar Momin Shah-Khan, MD
West Virginia University, Department of Medicine. Morgantown, WV

Anshu Mahajan, MD
West Virginia University, Section of Digestive Diseases. Morgantown, WV

Corresponding Author: Fahad Chaudhary MD, Department of Internal Medicine, 1 Medical Center Drive, Morgantown, WV 26505. Email: fahad.chaudhary@hsc.wvu.edu

Conflicts of Interest: The authors have no financial conflicts of interest.

Informed Consent: Obtained from patient’s son who is the medical power of attorney, as the patient is deceased.

Abstract
Choroidal melanoma is a rare and deadly ocular cancer. We report a case of metastatic choroidal melanoma presenting as isolated gastric varices (IGV1). A 69-year old female with a history of choroidal melanoma treated with plaque radiotherapy presented with a 3 month history of epigastric pain, especially after spicy meals. EGD revealed isolated gastric varices. Initial right upper quadrant ultrasound was negative, as was a liver biopsy. CT Abdomen and Pelvis with intravenous contrast obtained for evaluation of splenic vein thrombosis incidentally revealed a right hepatic lobe mass. Repeat biopsy of the liver mass confirmed a diagnosis of metastatic melanoma. Additional imaging revealed brain metastasis. The patient eventually chose palliative care with hospice. To our knowledge, this is the first reported case of metastatic choroidal melanoma presenting as isolated gastric varices.

Introduction
Uveal melanoma is rare, occurring in 5 cases per million person years.1 It is the most common primary ocular cancer.2 95% of all uveal melanomas arise from the choroid and are referred to as choroidal melanoma.3 About half of patients don’t present with symptoms while the other half can present with flashes, floaters and visual field disturbances. Although local treatment with radiation therapy or enucleation prevents local recurrence in 95% of cases, nearly 50% of patients are at risk for metastatic disease.4 The most common sites of metastatic disease are to the liver, lungs and brain.5 We report a case of choroidal melanoma metastasis to the liver presenting as isolated gastric varices. We will discuss management and screening of patients with a history of choroidal melanoma.

Case Presentation
A 69 year old female with a history choroidal melanoma treated in 2012 with plaque brachytherapy, hypothyroidism, anxiety and tobacco use (1/2 packs per day for 40 years) was referred for outpatient esophagogastroduodenoscopy (EGD) in 2015 when she presented to her primary care provider with a 12 week history of epigastric abdominal pain. Patient’s pain had been worsening from a dull ache to sharp sensations, usually worsening at night and after meals. Patient had been prescribed Omeprazole 40 mg daily which only provided minimal relief. She denied alarm symptoms such as a history of anemia, gastrointestinal bleeding or unintentional weight loss. She also denied a history of alcohol or illicit drug use.

On the day of the procedure, vital signs were unremarkable. Physical examination revealed a well-nourished, elderly female with mild tenderness to palpation in her epigastrium with no rebound or guarding. EGD revealed a normal esophagus, small hiatal hernia, mild antral gastritis, normal duodenum, but interestingly, an isolated fundal gastric varices (IGV1) on retroflexion (Figure 1). The patient previously had serially normal liver function tests for surveillance due to her history of Choroidal melanoma and potential risk for liver metastasis. Additionally, she had also had serial magnetic retrograde cholangiopancreatography (MRCP) studies which revealed mild fatty changes of the liver and small lesions of early arterial enhancement in bilateral hepatic lobes thought to be hemangiomas, based on their stability over 3 years. Most recent MRCP had been 6 months prior to EGD. No imaging studies had been consistent with signs of cirrhosis. Furthermore, laboratory testing for chronic liver diseases was also negative.

We subsequently turned our evaluation for an etiology of IGV1 to searching for a splenic vein thrombosis. Right upper quadrant ultrasound and a specific splenic vein ultrasound were unremarkable. CT abdomen and pelvis with intravenous contrast confirmed negative testing for splenic vein thrombosis but incidentally revealed interval development of low attenuation mass in the right hepatic lobe with nodular margins and no evidence of central filling on delayed imaging. MRCP revealed a 5.6 cm x 3.5 cm x 6.1 cm, peripheral right hepatic lobe mass, as well as multiple smaller arterial enhancing
masses throughout the right lobe of the liver with overall findings suspicious for metastatic disease. Subsequently, the patient underwent CT guided percutaneous core needle biopsy of the right hepatic lobe mass. Imaging also revealed likely lymph node involvement in the gastro-hepatic region with largest lymph node measuring 1.5 cm.

Biopsy of the liver mass demonstrated a spindle and epithelioid cell malignant neoplasm in background of liver tissue. Traces of pigment suggestive of melanin were noted (Figure 1). Immunohistochemical stains were performed and the tumor cells were positive for MART-1 and S-100, while negative for pancytokeratin. Clinical history with morphology and immunophenotype on biopsy supported diagnosis of metastatic melanoma. Subsequent staging also revealed patient had brain metastases in the left occipital lobe. Patient eventually underwent palliative therapy with Nivolumab. Patient went into hospice care and died 7 months later.

Discussion

To the best of our knowledge, this is the first case report which identified metastatic choroidal melanoma as the etiology for findings of IGV1 on upper endoscopy. Metastasis to the liver is the most common (93%) followed by the lung (24%) and bones (16%). There have been 3 other case reports where liver metastasis were found at the time of diagnosis of choroidal melanoma as well as case reports of patients found to have liver metastasis 9 years after initial treatment of local disease. Studies have shown that up to half of all patients diagnosed with choroidal melanoma are at risk for metastatic disease. Choroidal melanoma has the propensity for early micro-metastasis followed by variable latency periods.

Anytime a patient presents with isolated gastric varix, this should prompt a workup for splenic vein occlusion/thrombus as these are the result of IGV1 in nearly 80% of cases. Splenic vein thrombosis results from injury to the splenic vein which can be attributed to acute pancreatitis, pancreatic neoplasms or trauma in the left upper quadrant. The patient in our case report was not found to have findings of splenic vein occlusion on multiple imaging modalities including ultrasound and CT scan. The other etiology for IGV1 can be attributed to portal hypertension.

This can obviously occur in patients with metastatic neoplasms to the liver. As previously described in studies of cancers in the liver, metastatic choroidal melanoma to the liver causes an increase in intrahepatic portal pressure. This in turn can cause portal hypertensive changes in the intestinal tract including IGV1.

Given the rarity of this melanoma, there are no consensus guidelines for screening patients who have a history of choroidal melanoma. In 2004, the Collaborative Ocular Melanoma Study (COMS), the largest studying uveal melanomas, recommended that after initial evaluation patients should undergo surveillance at least annually with a suggestion of every 6 months for the first 5 years and then annually afterwards.

Imaging studies utilized for surveillance include ultrasound, CT and MRI of the liver. Ultrasound alone has been found to be specific (100%) but not nearly sensitive enough. MRI has shown to have better sensitivity (95%) with similar specificity. It should be noted that recently there has been a debate as to whether screening with bi-annual MRI is economically practical given that there is a lack of evidence in improvement of overall survival even with screening. However, as will be discussed later, there is promising...
research into hepatic metastasis resection (although not specifically uveal melanoma). Nonetheless, the authors recommend screening at time of diagnosis of choroidal melanoma as well as every 6 months for the first 5 years and then annually for prognostication and patient planning for end of life diagnosis. We suggest a modified screening approach outlined in JAMA Ophthalmology, where patients receive right upper quadrant ultrasound every 6 months with subsequent MRI, if needed, to investigate abnormal findings bi-annually for 5 years and annually afterwards.7 Screening at this interval has been shown to discover 93% of metastasis prior to patient symptoms. Limited studies have also revealed that contrast-enhanced ultrasound shows comparable sensitivity to MRI and may also be an affordable screening test in the outpatient setting.10

Median survival of patients with choroidal melanoma is 45 months and 2-9 months for patients diagnosed with liver metastasis.11 Overall survival has been unchanged since the early 1970s.12 Currently, there is no standard of care for treatment of metastatic disease. In patients with liver metastasis, survival has shown to be better in those that undergo resection compared to those that do not, however, it is unclear if this is due to selection bias with surgery typically attempted more often in younger patients.13 More study is needed in the treatment of metastatic choroidal melanoma to not only improve overall survival but better identify populations who are candidates for metastatic resection.

Conclusion

We report a unique presentation of rare choroidal melanoma that involved metastasis to the liver. These patients require indefinite screening for metastasis given the latency of the disease with no current standard of care in treating metastatic disease. However, promising sensitivity of contrast-enhanced ultrasound and metastatic resection may yield improvements in survival not seen in the past 45 years.

References