Retroperitoneal Tumor: A Case Report

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Abstract

Neurofibromas are common benign neoplasms that are composed of various components, including nerve fibers, fibroblasts, collagen, and supporting tissues and cells. They are commonly known as benign growths composed of a bundle of nerve fibers and are commonly associated with patients suffering from Neurofibromatosis. [3][4] Retroperitoneal masses are also a common incidental finding in a large proportion of the population. These masses are commonly found on routine abdominal CT scans in patients who are suffering from other conditions or symptoms that require this type of imaging. [5]

This case report deals with a large retroperitoneal mass, measuring 10.9 x 7.0 x 12.9 cm as well as the concurrent management of appendicitis in a 34 year old male patient. This report includes the patients course of treatment, including history and physical examination, diagnostic reports, CT scan, PET scan and the surgical and pathology reports of a lymph node biopsy.

Abdominal and back pain can occur in a multitude of situations. Retroperitoneal masses are typically not the initial diagnosis one would suspect. This case describes the various diagnostic processes to manage such findings. It also divulges the importance of further testing in these types of situations as not all retroperitoneal masses are benign and asymptomatic.

Introduction

Retroperitoneal masses are a common benign finding in a variety of patient populations. Neurofibromas in particular are complex tumors that are a “mix of Schwann cells, perineural-like cells, and fibroblasts, interspersed with nerve fibers, wire-like strands of collagen, and myxoid matrix.” [3] There is usually no known cause as to why neurofibromas form. [6]

Most patient’s with isolated neurofibromas (90% of cases) present between the ages of 20-30 years old. A characteristic finding is “the presence of intratumor nerve fibers”. [3] Over time these tumors may potentially grow in size and present with numbness, or pain of a particular area due to the mass compressing nearby nerves or growing within nerve sheaths. [6]

Classically neurofibromas can present as cutaneous or intraneural lesions.

Cutaneous neurofibromas are typically small tumors, that are capable of causing minor pain or bleeding locally, however they do not typically cause neurologic discrepancies. Intraneural neurofibromas are typically found to be “deeper, focal, well-circumscribed, fusiform lesions involving nerve roots, nerve trunks, nerve plexuses, or peripheral nerves.”[3] The characteristic presentation of these tumors is the finding of a mass with localized pain, or some deficits within sensorimotor neurologic processes. [3] The involvement of several nerve bundles within a single neurofibroma is termed a plexiform neurofibroma. [6]

Solitary neurofibromas represent 2/3 of the neurofibromas that are excised during the time of surgery. These tumors very rarely become malignant. The remaining 1/3 represent neurofibromas that are seen in patients with neurofibromatosis type 1. Therefore, patient’s who are found to have multiple neurofibromas are typically diagnosed with neurofibromatosis type 1. [3]

Solitary neurofibromas are typically managed through observation or surgical excision. If the tumor presents in a location that is difficult to reach or is not invading other organs or nerve structures, the clinician will typically just observe the mass for changes as time progresses. However, if the location and size of the tumor are no longer manageable, or the patient is experiencing debilitating symptoms, surgical excision may be required. The goal of surgical excision is to remove the tumor without negatively disrupting the surrounding neurologic processes. [3][6]

Case Presentation

This report involves a 34-year-old African American male with no significant past medical history who presents with acute abdominal pain to the emergency department. The patient was also experiencing nausea, vomiting, and right lower quadrant pain. Physical examination showed signs of appendicitis and revealed multiple enlarged lymph nodes. A CT scan of the abdomen and pelvis was obtained following the administration of oral and intravenous contrast. The imaging studies demonstrated an enlarged, fluid filled appendix that measured 1.3 cm in diameter, which further confirmed the suspicions of appendicitis. The CT scan also revealed the presence of a retroperitoneal multi-septated mass on the left side of the retroperitoneum. The mass contained focal calcifications and displaced the left kidney anteriorly. The mass measured 10.9 x 7.0 x 12.9 cm. The CT scan also confirmed the enlargement of several lymph nodes; including the para-aortic, external iliac, and bilateral inguinal lymph nodes. Multiple tiny nodules were noted at the lung bases as well as a 0.9 x 0.9 cm soft tissue density noted at the posterior aspect of the L2 vertebral body, which raised suspicions for metastatic disease.

Biopsy of the mass performed during appendectomy, resulted in the diagnosis of a benign neurofibroma. Further diagnostics were performed due to a yearlong history of back pain, fatigue, and a 15-20 pound weight loss. These studies revealed that the patient was infected with Human Immunodeficiency Virus (HIV).

A PET scan was ordered to further investigate the nodules found in the lungs and the soft tissue findings of the L2 vertebra. The PET scan found multiple lymph nodes with slightly increased metabolic activity in the posterior cervical, subclavicular, and axillary areas. Miliary-type diffuse bilateral pulmonary nodules were also present. The retroperitoneal mass itself had no significant increased metabolic activity. However, there were enlarged bilateral pelvic and inguinal lymph nodes that were mildly metabolically active. The conclusion of the CT scan demonstrated that the findings were moderately suspicious for malignancy.

A fine needle aspiration of the left supraclavicular lymph node was later ordered and the results came back as low cellularity and negative for Hodgkin's cells.

Discussion

The majority of diagnosed neurofibromas are benign and in 90% of cases these neurofibromas are solitary and isolated lesions. [3] The size and number of tumors, as well as its origin play an important role in determining whether one is dealing with an individual collection of cells, or whether one is dealing with metastatic disease. Individual tumors are typically found in one area and on one side of the body and tend to be larger. [6] Therefore the fact that this patient is dealing with a right sided, large retroperitoneal mass is suggestive of a benign underlying etiology. The large size of neurofibromas can lead to the compression of surrounding structures. The compression of nerves can lead to pain and numbness, whereas the compression of larger surrounding structures can lead to more serious complications ranging from ureteral obstruction, lower back pain, hydronephrosis, lower extremity swelling and discoloration and bowel obstruction.

The initial diagnosis of neurofibromas varies. Typically these structures can be found as an incidental finding. In this case the patient presented due to appendicitis, and was subsequently diagnosed with an abdominal CT scan and biopsy. Further diagnostic studies such as PET scans and lymph node biopsies were mandated in this case due to the finding of enlarged lymph nodes as well as multiple nodules concentrated in the base of the lungs, with some small nodules located within the bilateral upper and middle lobes. The patient also had various blood draws to determine if there were any inflammatory marker abnormalities, which all came back negative.

Management of neurofibromas depends on a variety of different factors including a patients age, current and past medical history, any underlying symptoms associated with the mass, the size of the tumor as well as any family history of similar masses. Typically for incidental findings, it is recommended to watch and wait. If the neurofibroma is causing the patient any form of discomfort or other physiologic symptoms, it is likely recommended to resect the mass. [3]

In patients with neurofibromatosis type 1, cutaneous neurofibromas are typically found in numerous amounts and can cover the majority of the patients body. 95% of patients with neurofibromatosis type 1 present with cutaneous forms and are typically the most bothersome to the patients. The best medical management for cutaneous

neurofibromas is surgical resection due to the fact that medical therapies have proven unsuccessful. [1] Other common therapies for small cutaneous neurofibromas include CO2 laser, modified biopsy removal, photocoagulation, electrodessication, and radiofrequency ablation. [1]

This patient presented with a history of nausea, vomiting and right lower quadrant pain, which contributed to his findings of appendicitis. The patient also had ongoing symptoms of weight loss and fatigue, which was determined to be due to his positive HIV status. The patient was put onto antiretrovirals for his HIV positivity and began to gain weight and decrease his viral load.

The inconclusive findings on this patients PET scan and lymph node biopsy lead to a conclusion of a benign retroperitoneal neurofibroma. There was no conclusive evidence of metastatic disease found on further biopsy, therefore the patient will continue to follow-up with his surgeon to monitor the progression and stability of the neurofibroma.

Conclusions

Benign neurofibromas are common findings in a multitude of people, varying from different genders, ages, and previous medical conditions. Neurofibromas, a collection of Schwann cells, myxoid matrix, nerve sheathes, fibroblasts and perineural cells are complex tumors. [3]

If the mass continues to grow on future abdominal imaging studies, then surgical resection and further testing should be considered. It is of utmost importance to take into consideration a patients symptoms in these particular cases due to the fact that even mild symptoms can potentially be related to the growth and invasion of the neurofibroma into surrounding structures. This case highlights the benign and incidental findings of a large retroperitoneal neurofibroma found in a patient with other acute and chronic medical conditions. These conditions were an important aspect of this patients diagnostic workup and further management.

References

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